Early diagnosis and treatment of NMOSD: Practical insights

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A conversation between:

Prof. John Chen

Mayo Clinic, Rochester,
MN, USA

Prof. Friedemann Paul

Charité - Universitätsmedizin Berlin,
Campus Mitte,
Germany

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Agenda

- Understanding the clinical features and presenting symptoms of NMOSD
- Initial assessment and differential diagnosis of NMOSD
- Early management of NMOSD to mitigate symptoms and reduce the risk of further attacks

Understanding the clinical features and presenting symptoms of NMOSD

Prof. John Chen

Mayo Clinic, Rochester,
MN, USA

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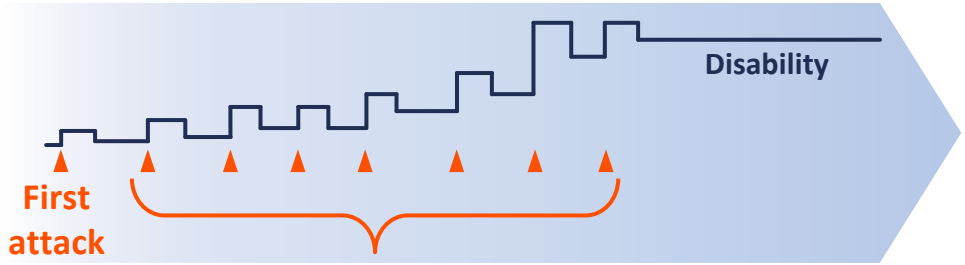
Clinical features of NMOSD

Clinical hallmarks:[1]

- Acute optic neuritis
- Transverse myelitis
- Area postrema syndrome

Course of disease:[1,2]

- A series of **discrete attacks**
- Recovery after an attack is often partial
- **Disability increases with each relapse**



Relapses[1]

- Occur in **80%–90% of patients**
- Frequently **within 3 years** after the initial episode

A definite diagnosis of NMOSD is essential to promptly and effectively counteract acute attacks and to prevent future attacks by initiating immunotherapy[3]

NMOSD, neuromyelitis optica spectrum disorder.

1. Oh J and Levy M. *Neurol Res Int.* 2012;2012:460825; 2. Chan K-H and Lee C-Y. *Int J Mol Sci.* 2021;22:8638; 3. Kümpfel T, et al. *J Neurol.* 2024;271:141–76.

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Initial assessment and differential diagnosis of NMOSD

Prof. John Chen

Mayo Clinic, Rochester,
MN, USA

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NMOSD diagnostic criteria algorithm[1]

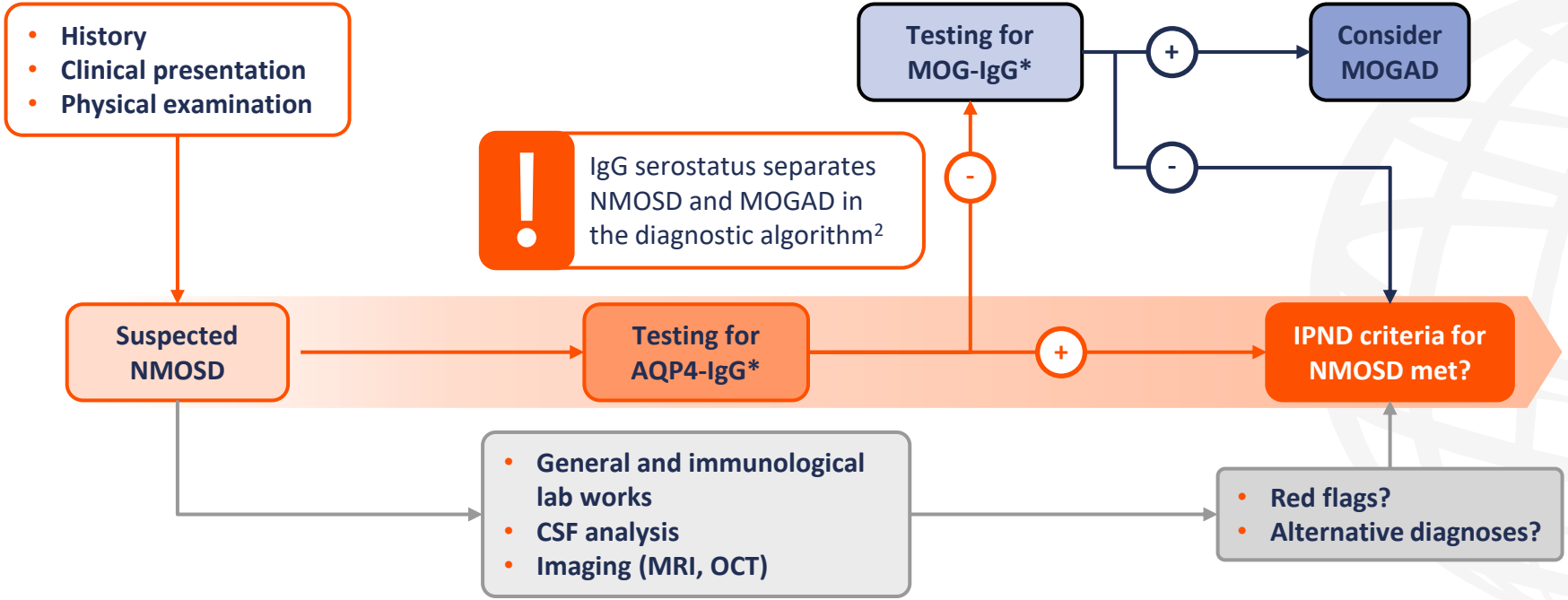

Figure adapted from Jarius S, et al. *J Neurol*. 2023;270:3341–68 (CC BY 4.0 www.creativecommons.org/licenses/by/4.0/).
*Tests should be repeated upon negative results.
AQP4, aquaporin-4; CSF, cerebrospinal fluid; IgG, immunoglobulin G; IPND, International Panel for Neuromyelitis Optica Diagnosis; MOG, myelin oligodendrocyte glycoprotein; MOGAD, myelin oligodendrocyte glycoprotein antibody-associated disease; MRI, magnetic resonance imaging; NMOSD, neuromyelitis optica spectrum disorder; OCT, optic coherence tomography.
1. Jarius S, et al. *J Neurol.* 2023;270:3341–68; 2. Cacciaguerra L, Flanagan EP. *Neurol Clin*. 2024;42:77–114.

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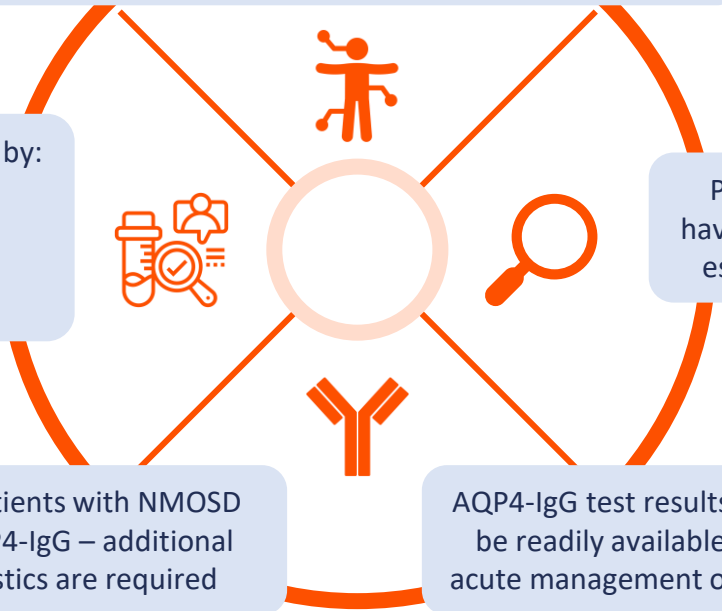
NMOSD diagnostic challenges

Diverse diseases with autoimmune, vascular, infectious, or neoplastic aetiologies can mimic these phenotypes of NMOSD

AQP4-IgG test results can be affected by:
- Assay methods (ideally CBA)
- Serologic status
- Disease stages
- Treatment types



Patients with NMOSD may only have limited clinical manifestations, especially in early disease stages

Some patients with NMOSD lack AQP4-IgG – additional diagnostics are required

AQP4-IgG test results may not be readily available for the acute management of NMOSD

AQP4-IgG, aquaporin-4 immunoglobulin G; CBA, cell-based assay; NMOSD, neuromyelitis optica spectrum disorder.

Kim S-M, et al. *Ther Adv Neurol Disord.* 2017;10:265–89.

Early management of NMOSD to mitigate symptoms and reduce the risk of further attacks

Prof. John Chen

Mayo Clinic, Rochester,
MN, USA

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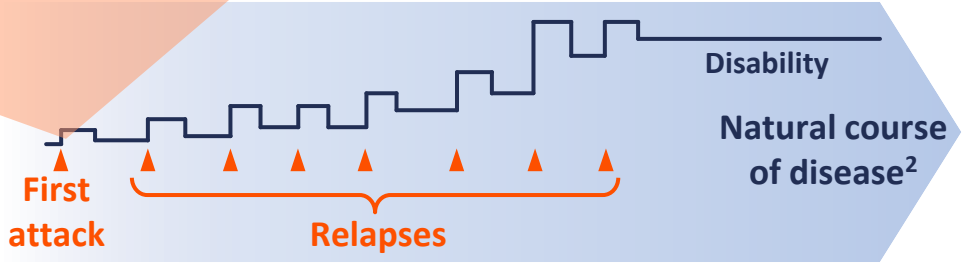
Treatment of NMOSD: Acute attacks

- Counteract the attack[1]
- Improve recovery[1]



IV methylprednisolone[1,3]
Traditionally used as first-line treatment

Immunoadsorption[1,3]
Alternative apheresis therapy if PLEX is contraindicated or unavailable

Plasmapheresis (PLEX)[1,3]
In addition to IV steroids or when treatment with IV steroids has failed

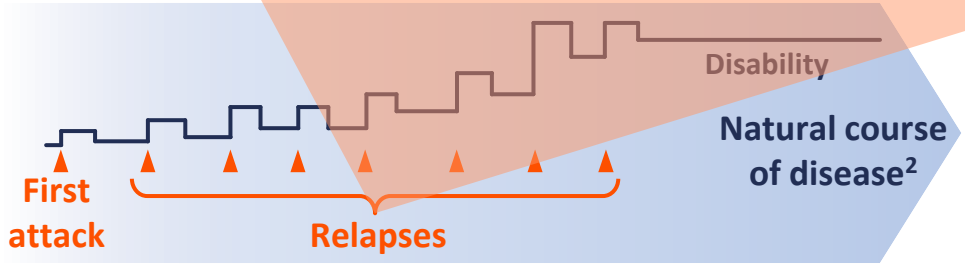
IV immunoglobulins[3]
Considered when IV steroids and apheresis are contraindicated

IV, intravenous; NMOSD, neuromyelitis optica spectrum disorder; PLEX, plasmapheresis.

1. Kümpfel T, et al. *J Neurol.* 2024;271:141–76; 2. Oh J and Levy M. *Neurol Res Int.* 2012;2012:460825; 3. Chan K-H and Lee C-Y. *Int J Mol Sci.* 2021;22:8638.

Treatment of NMOSD: Long-term maintenance

- Reduce the risk of further attacks[1]
- Prevent the accumulation of disability[1]



Eculizumab[3,4]	Inebilizumab[5,6]	Ravulizumab[7,8]	Satralizumab[9,10]	Rituximab[1]
Anti-C5 complement protein mAb	Anti-CD19 mAb	Anti-C5 complement protein mAb	Anti-IL-6 receptor mAb	Anti-CD20 mAb

Approved in Europe and the USA for the treatment of adult patients* with NMOSD who test positive for AQP4-IgG[3–10]

Approved in Japan; used off-label in many countries[1]

*Satralizumab is also EMA-approved in adolescent patients from 12 years of age.

AQP4-IgG, aquaporin-4 immunoglobulin G; IL, interleukin; mAb, monoclonal antibody; NMOSD, neuromyelitis optica spectrum disorder.

1. Kümpfel T, et al. *J Neurol.* 2024;271:141–76; 2. Oh J and Levy M. *Neurol Res Int.* 2012;2012:460825; 3. FDA. Eculizumab PI. 2024; 4. EMA. Eculizumab SmPC. 2023; 5. FDA. Inebilizumab PI. 2020; 6. EMA. Inebilizumab SPC. 2024; 7. FDA. Ravulizumab PI. 2024; 8. EMA. Ravulizumab SPC. 2023; 9. FDA. Satralizumab PI. 2022; 10. EMA. Satralizumab SmPC. 2023. All PIs available at: www.accessdata.fda.gov/scripts/cder/daf/index.cfm. All SPCs available at: www.ema.europa.eu/en/medicines. All accessed 16 May 2024.

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