Early diagnosis and treatment of NMOSD: Practical insights

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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

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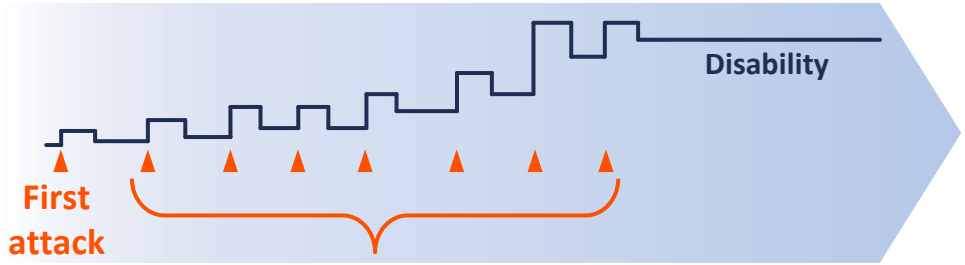
Clinical features of NMOSD

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.

Initial assessment and differential diagnosis of NMOSD

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NMOSD diagnostic criteria algorithm[1]

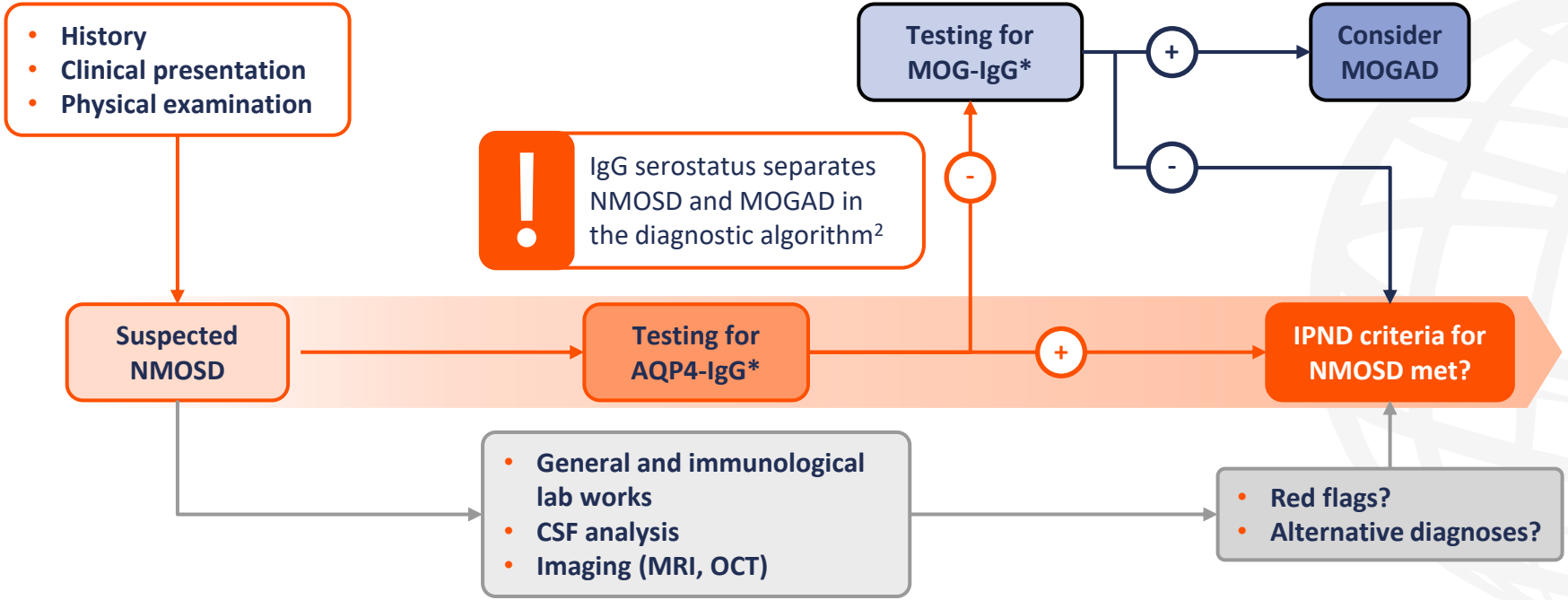

Figure adapted from Jarius S, et al. *J Neurol*. 2023;270:3341–68 (CC BY 4.0 www.creativecommons.org/licenses/by/4.0/).
*Tests should be repeated upon negative results.
AQP4, aquaporin-4; CSF, cerebrospinal fluid; IgG, immunoglobulin G; IPND, International Panel for Neuromyelitis Optica Diagnosis; MOG, 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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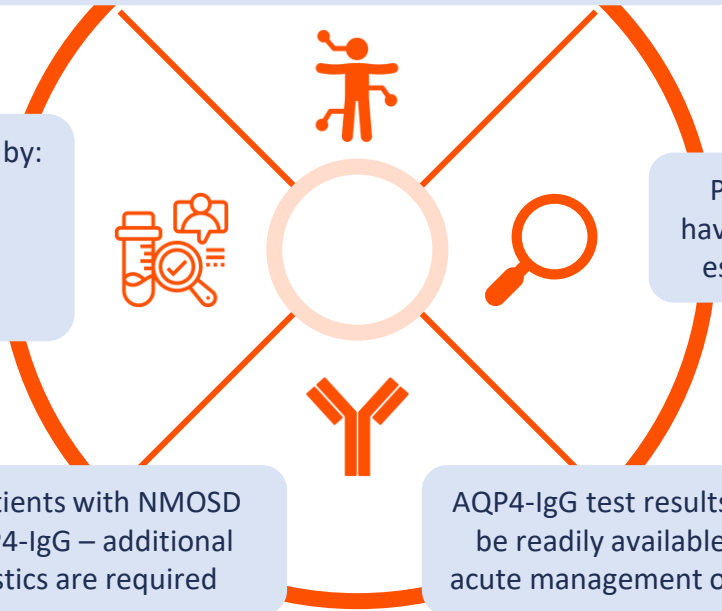
NMOSD diagnostic challenges

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.

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

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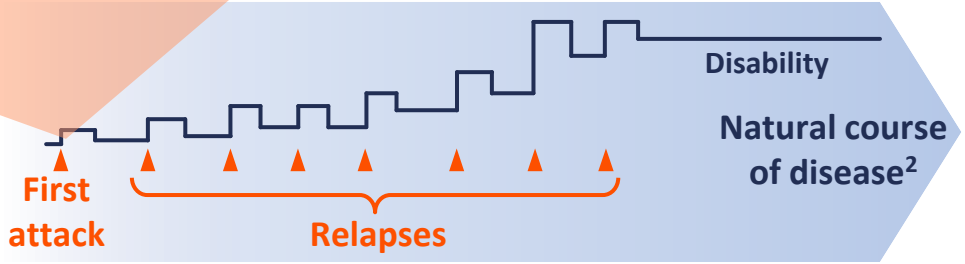
Treatment of NMOSD: Acute attacks

- 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.

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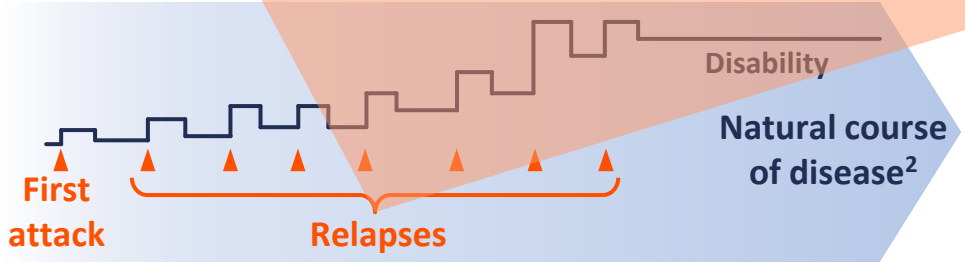
Treatment of NMOSD: Long-term maintenance

- 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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