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Early diagnosis and treatment of NMOSD: Practical insights

Practice aid for neuromyelitis optica spectrum disorder

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NMOSD clinical features

Clinical hallmarks:[1,2]

- Acute optic neuritis
- Transverse myelitis
- Area postrema syndrome (nausea, vomiting, hiccups)

Course of disease:[1,3]

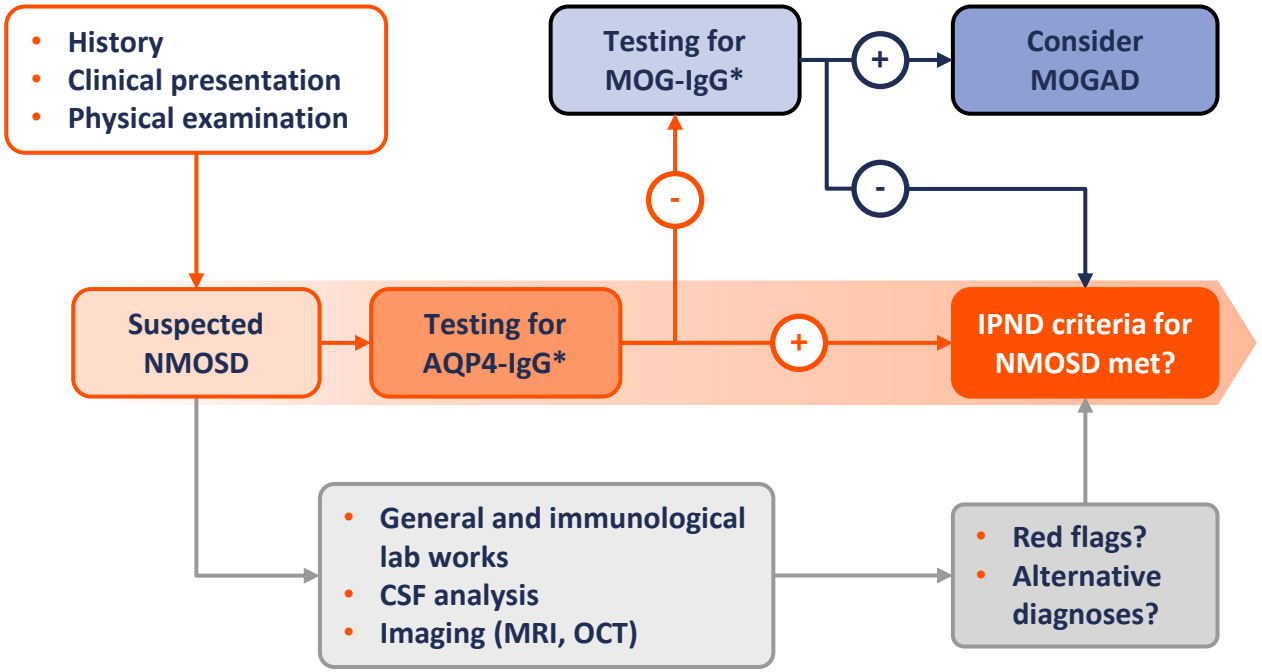
- A series of **discrete attacks**
- Recovery after an attack is often partial
- **Disability increases with each relapse**

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[4]

NMOSD diagnostic criteria algorithm[5]



IgG serostatus separates NMOSD and MOGAD in the diagnostic algorithm[6]

- **CBA is the optimal, recommended test for both AQP4-IgG and MOG-IgG[5]**
- Other assays, such as IHC and ELISA, are less sensitive and/or specific than CBA[5]
- IHC or ELISA can be used for detecting AQP4-IgG if CBA is not available[5]
  - A CBA should be striven for and performed as soon as it becomes available[5]

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.

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Treatment of NMOSD

IV methylprednisolone[3,4]
Traditionally used as first-line treatment

Plasmapheresis (PLEX) [3,4]
In addition to IV steroids or when IV steroids have failed

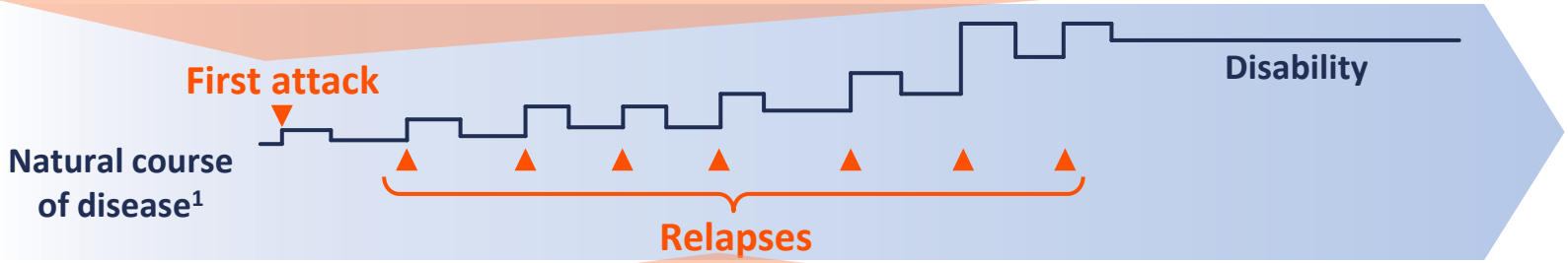
Immunoadsorption[3,4]
Alternative apheresis if PLEX is contraindicated or unavailable

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

Acute attacks

- Counteract the attack[4]
- Improve recovery[4]

For patients with severe attacks, simultaneous treatment with glucocorticoids and apheresis should be considered[4]



Long-term maintenance

- Reduce the risk of further attacks[4]
- Prevent the accumulation of disability[4]

Eculizumab[7,8]
Anti-C5 complement protein mAb

Inebilizumab[9,10]
Anti-CD19 mAb

Ravulizumab[11,12]
Anti-C5 complement protein mAb

Satralizumab[13,14]
Anti-IL-6 receptor mAb

Rituximab[4]
Anti-CD20 mAb

Approved in Europe and the USA for the treatment of adult patients* with NMOSD who test positive for AQP4-IgG[7–14]

Approved in Japan; used off-label in many countries[4]

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

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Abbreviations and references

Abbreviations

AQP4-IgG, aquaporin-4 immunoglobulin G; CBA, cell-based assay; CSF, cerebrospinal fluid; ELISA, enzyme-linked immunosorbent assay; EMA, European Medicines Agency; FDA, Food and Drug Administration; IHC, immunohistochemistry; IL, interleukin; IPND, International Panel for Neuromyelitis Optica Diagnosis; IV, intravenous; mAb, monoclonal antibody; MOG-IgG, myelin oligodendrocyte glycoprotein immunoglobulin G; MOGAD, myelin oligodendrocyte glycoprotein antibody-associated disease; MRI, magnetic resonance imaging; NMOSD, neuromyelitis optica spectrum disorder; OCT, optic coherence tomography; PLEX, plasmapheresis.

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