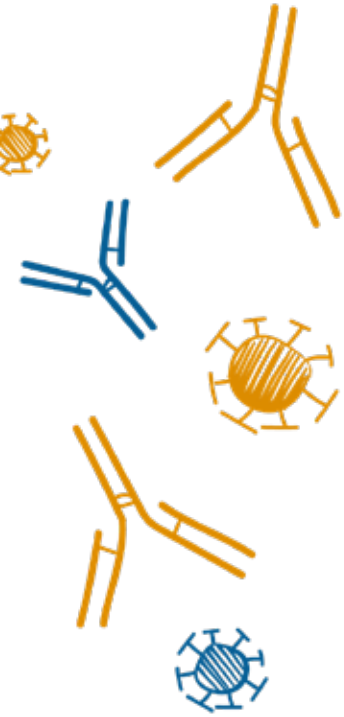
NMOWN THE PATH FORWARD

# A PRIMER FOR YOUR PATH FORWARD

**Neuromyelitis optica spectrum disorder (NMOSD) is a rare and unpredictable disease. The condition is caused by inflammation in the central nervous system, which is made up of the brain, optic nerve, and spinal cord.[1,2]**

## Who NMOSD Affects

- The median age of onset is **40 years** but can range from age 2 to 85[3]
- **Women may be 9 times more likely** to be impacted versus men[4]
- There is a **2-to-3-fold higher prevalence in Asian and Black** populations[4,5]
- An estimated **10,000-15,000 people** are diagnosed in the United States[5]



## NMOSD and the Immune System

NMOSD is an autoimmune disease of the central nervous system, although the exact cause of the disease is unknown.[6] NMOSD relapses (sometimes referred to as attacks) occur when the body's immune system mistakenly attacks healthy cells.[7]

Your doctor will test you for specific immune system proteins called antibodies to aid in diagnosis and to help determine treatment options.[7,8]

- **Aquaporin-4 (AQP4)**

  80% of NMOSD cases are considered "AQP4 seropositive" and involve AQP4, a common protein responsible for helping water enter and leave nervous system cells[7]

- **Other Types**

  Less commonly, NMOSD is diagnosed without anti-AQP4 antibody involvement, although antibodies against other cellular proteins can sometimes be found[8]

## Permanent Disability Can Result From Relapses

Individuals with NMOSD often do not fully recover from relapses. Without treatment, severe and permanent disability may result from accumulating damage caused by inflammation.[2,9,10]

- Relapses can cause permanent vision impairment, muscle weakness, paralysis, pain, and fatigue[11-13]
  - **Within 3 years**
    **69%** of patients have severe vision loss in at least one eye[10]
  - **After about 6 years**
    Up to **18%** of patients lose vision in both eyes, and **34%** may have a permanent motor disability[11]
- NMOSD disease progression, severity, and relapse risks are unpredictable and vary greatly among patients[14]


## Misdiagnosis: NMOSD vs MS[15,16]

41% of patients with NMOSD have reported an initial misdiagnosis of multiple sclerosis (MS). Doctors may be more familiar with MS, and the conditions can appear similar at first look. However, several factors differentiate the diseases:

| | | |
|---|---|---|
| **Key differentiating symptoms** | In NMOSD, **severe vision impairment** is more common | In MS, cognitive and psychological symptoms like **memory loss or depression** are common |
| **Differences in relapses** | In NMOSD, **relapses are more severe** and may lead to an accumulation of **irreversible damage and disability** in affected areas | In MS, individual episodes are mild and over time may or may not lead to **progressive disability** |
| **Biomarkers** | In NMOSD, a blood test will come back as **positive for Aquaporin-4 (AQP4) or MOG** antibodies | In MS, a biomarker test will always come back **negative for AQP4** antibodies. MOG antibodies are not found in "classical" MS and could be indicative of another condition |

Because NMOSD relapses can lead to permanent disability, **it's important to get an early and definitive diagnosis and start on a treatment.**

**References: 1** Ajmera MR, Boscoe A, Mauskopf J, Candrilli SD, Levy M. Evaluation of comorbidities and health care resource use among patients with highly active neuromyelitis optica. *J Neurol Sci*. 2018;384:96-103. **2** What Is NMO? Guthyjacksonfoundation.org. Accessed January 12, 2021 http://www.guthyjacksonfoundation.org/neuromyelitis-optica-nmo. **3** Mealy MA, Wingerchuk DM, Greenberg BM, Levy M. Epidemiology of neuromyelitis optica in the United States. *Arch Neurol*. 2012;69(9):1176-1180. **4** Wingerchuk DM. Neuromyelitis optica: effect of gender. *J Neurol Sci*. 2009;286(1-2):18-23. **5** Flanagan EP, et al. Epidemiology of aquaporin-4 autoimmunity and neuromyelitis optica spectrum. *Ann Neurol*. 2016;79(5):775-783. **6** Neuromyelitis optica. Mayoclinic.org. https://www.mayoclinic.org/diseases-conditions/neuromyelitis-optica/symptoms-causes/syc-20375652 Published December 15, 2015. Accessed January 12, 2021. **7** Layman's Guide to NMO. Sumairafoundation.org. Accessed January 12, 2021 https://www.sumairafoundation.org/laymans-guide-to-nmo. **8** Lana-Peixoto MA, Talim N. Neuromyelitis optica spectrum disorder and anti-MOG syndromes. *Biomedicines*. 7(2);42. **9** Kimbrough DJ, et al. Treatment of neuromyelitis optica: review and recommendations. *Mult Scler Relat Disord*. 2012;1(4):180-187. **10** Baranello RJ, Avasarala JR. Neuromyelitis optica spectrum disorders with and without aquaporin 4 antibody: characterization, differential diagnosis, and recent advances. *J Neuro Ther*. 2015;1(1):9-14. **11** Kitley J, et al. Prognostic factors and disease course in aquaporin-4 antibody-positive patients with neuromyelitis optica spectrum disorder from the United Kingdom and Japan. *Brain*. 2012;135(6):1834-1849. **12** Cabra P, et al. Relapsing neuromyelitis optica: long term history and clinical predictors of death. *J Neurol Neurosurg Psychiatry*. 2009;80:1162-1164. **13** Eaneff S, et al. Patient perspectives on neuromyelitis optica spectrum disorders: data fromthe PatientsLikeMe online community. *Mult Scler Relat Disord*. 2017;17:116-122. **14** Kunchok A, et al. Clinical and therapeutic predictors of disease outcomes in AQP4-IgG+ neuromyelitis optica spectrum disorder. *Mult Scler Relat Disord*. 2020;38:101868-101876. **15** Beekman J, et al. Neuromyelitis optica spectrum disorder: patient experience and quality of life. *Neurol Neuroimmunol Neuroinflamm*. 2019;6(4). **16** Siegel Rare Neuroimmune Association (SRNA). Neuromyelitis optica spectrum disorder. Wearesrna.org. https://wearesrna.org/living-with-myelitis/disease-information/neuromyelitis-optica-spectrum-disorder Accessed January 12, 2021. **17** Hamid SHM, et al. What proportion of AQP4-IgG-negative NMO spectrum disorder patients are MOG-IgG positive? a cross-sectional study of 132 patients. *J Neurol*. 2017;64(10):2088-2094.