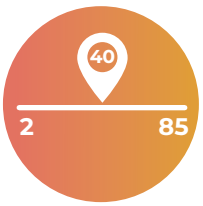
What Is NMOSD?

Neuromyelitis optica spectrum disorder (NMOSD) is a rare and debilitating autoimmune disease of the central nervous system characterized by immune-mediated damage to the optic nerve, brain stem and spinal cord.[1,2]



The median age of disease onset is **40** but can range from ages **2 to 85**.[3]

About **10,000 to 15,000** people in the United States have NMOSD.[4]

All people can be affected by NMOSD—but **women may be 9 times** more likely to be impacted than men.[5]

The prevalence is **2- to 3-fold higher in Black and Asian populations**.[4,5]

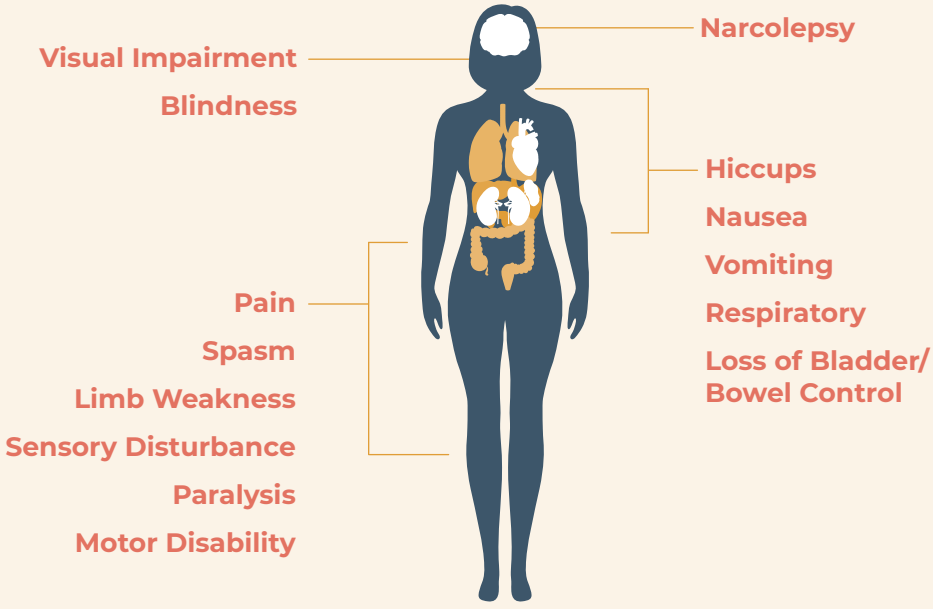
Role of B Cells in NMOSD

In NMOSD, damage is caused when CD19+-expressing B cell lymphocytes (plasmablasts and plasma cells) produce AQP4-IgG, triggering an escalating autoimmune reaction.[6]

Depletion of CD19+ B cells has an impact on the ability of the immune system to attack of AQP4 channels on astrocytes and other neurons.[6]

Symptoms of Attacks and Ongoing Disability

Individuals with NMOSD often do not fully recover from attacks, and permanent disability results from accumulating damage from attacks.[2,9,10]



NMOSD progression and severity **varies greatly** among patients, and attacks and disease severity are unpredicatble.[7]

Attacks may cause **permanent muscle weakness**, paralysis, pain and fatigue.[2]

Within **3 years**, **69%** of patients have severe vision loss in at least one eye. After about **6 years**, as many as **18%** of patients lose vision in both eyes and **34%** may have a permanent motor disability.[11, 12]

Science Behind NMOSD

The cause is **unknown** but NMOSD may manifest following an infection or may be associated with another autoimmune condition.[13]

In about **80%** of cases, antibodies to **aquaporin-4** (AQP4) are present ("seropositive cases"). AQP4 is an important water channel protein in the central nervous system (CNS).[1]

The cause in **seronegative cases** is less clear, but some patients have antibodies to myelin oligodendrocyte glycoprotein.[14, 15]

Misdiagnosis

41% of NMOSD patients have reported an initial misdiagnosis of multiple sclerosis (MS).[8, 16] However, several factors differentiate the diseases:

NMSOD	MS
Key symptom: severe vision impairment	Key symptoms: cognitive and psychological symptoms (eg, memory loss or depression)
Severe acute episodes can lead to permanent disability	Progressive disability caused by individual, typically mild, episodes
Permanent CNS damage	Often experience better recovery from attacks
AQP4 antibody seropositive*	AQP4 antibody seronegative

**80% of people with NMOSD are AQP4 antibody seropositive, but people with MS are always seronegative.*[15]

NMOSD and MS are treated in different ways, and early detection and treatment help ensure the best outcomes. Differential diagnosis of NMOSD and MS is important because MS treatments may be ineffective or may exacerbate NMOSD.[17,18]

Burden of NMOSD

Due to the cost of managing the underlying disease and the treatment of attacks, **NMOSD imposes a significant financial burden on patients**.[8] Hospitalizations and emergency department visits also contribute significantly to the economic burden of the disease.[1]

60% of patients report **significant impact on work, activities and being able to accomplish things** all or most of the time.[17]

55% of people with NMOSD report **fatigue**, while **54%** have some level of **cognitive impairment**. Bladder and bowel problems, along with sexual dysfunction, are also common.[17]

Despite the physical impairment and challenges of NMOSD, studies show that patients demonstrate psychological resilience.[8] This resilience, coupled with the changing NMOSD treatment landscape, may offer hope for the NMOSD community.

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