



NINDS Neuromyelitis Optica Information Page

Synonym(s): Devic's Syndrome

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What is Neuromyelitis Optica ?

Neuromyelitis optica (NMO) is an uncommon disease syndrome of the central nervous system (CNS) that affects the optic nerves and spinal cord. Individuals with NMO develop optic neuritis, which causes pain in the eye and vision loss, and transverse myelitis, which causes weakness, numbness, and sometimes paralysis of the arms and legs, along with sensory disturbances and loss of bladder and bowel control. NMO leads to loss of myelin, which is a fatty substance that surrounds nerve fibers and helps nerve signals move from cell to cell. The syndrome can also damage nerve fibers and leave areas of broken-down tissue. In the disease process of NMO, for reasons that aren't yet clear, immune system cells and antibodies attack and destroy myelin cells in the optic nerves and the spinal cord.

Historically, NMO was diagnosed in patients who experienced a rapid onset of blindness in one or both eyes, followed within days or weeks by varying degrees of paralysis in the arms and legs. In most cases, however, the interval between optic neuritis and transverse myelitis is significantly longer, sometimes as long as several years. After the initial attack, NMO follows an unpredictable course. Most individuals with the syndrome experience clusters of attacks months or years apart, followed by partial recovery during periods of remission. This relapsing form of NMO primarily affects women. The female to male ratio is greater than 4:1. Another form of NMO, in which an individual only has a single, severe attack extending over a month or two, is most likely a distinct disease that affects men and women with equal frequency. The onset of NMO varies from childhood to adulthood, with two peaks, one in childhood and the other in adults in their 40s.

In the past, NMO was considered to be a severe variant of multiple sclerosis (MS) because both can cause attacks of optic neuritis and myelitis. Recent discoveries, however, suggest it is a separate disease. NMO is different from MS in the severity of its attacks and its tendency to solely strike the optic nerves and spinal cord at the beginning of the disease. Symptoms outside of the optic nerves and spinal cord are rare, although certain symptoms, including uncontrollable vomiting and hiccups, are now recognized as relatively specific symptoms of NMO that are due to brainstem involvement.

The recent discovery of an antibody in the blood of individuals with NMO gives doctors a reliable biomarker to distinguish NMO from MS. The antibody, known as NMO-IgG, seems to be present in about 70 percent of those with NMO and is not found in people with MS or other similar conditions.

Is there any treatment?

There is no cure for NMO, but there are therapies to treat an attack while it is happening, to reduce symptoms, and to prevent relapses. Doctors usually treat an initial attack of NMO with a combination of a corticosteroid drug (methylprednisolone) to stop the attack, and an immunosuppressive drug (azathioprine) for prevention of subsequent attacks. If frequent relapses occur, some individuals may need to continue a low dose of steroids for longer periods. Plasma exchange (plasmapheresis) is a technique that separates antibodies out of the blood stream and is used with people who are unresponsive to corticosteroid therapy. Pain, stiffness, muscle spasms, and bladder and bowel control problems can be managed with the appropriate medications and therapies. Individuals with major disability will require the combined efforts of occupational therapists, physiotherapists, and social services professionals to address their complex rehabilitation needs.

What is the prognosis?

Most individuals with NMO have an unpredictable, relapsing course of disease with attacks occurring months or years apart. Disability is cumulative, the result of each attack damaging new areas of myelin. Some individuals are severely affected by NMO and can lose vision in both eyes and the use of their arms and legs. Most individuals experience a moderate degree of permanent limb weakness from myelitis. Muscle weakness can cause breathing difficulties and may require the use of artificial ventilation. The death of an individual with NMO is most often caused by respiratory complications from myelitis attacks.

What research is being done?

The National Institute of Neurological Disorders and Stroke (NINDS) conducts research related to NMO in laboratories at the NIH and also supports additional research through grants to major medical institutions across the country. Much of this research focuses on finding better ways to prevent, treat, and ultimately cure rare neurological syndromes such as NMO.

NIH Patient Recruitment for Neuromyelitis Optica Clinical Trials

- ▶ [At NIH Clinical Center](#)
- ▶ [Throughout the U.S. and Worldwide](#)

Organizations

Multiple Sclerosis Foundation

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National Eye Institute (NEI)

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National Organization for Rare Disorders (NORD)

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