Neuromyelitis Optica (NMO)

What is NMO?

Neuromyelitis optica (NMO), sometimes known as 'Devic's disease' is a rare autoimmune disorder of the central nervous system (CNS) where antibodies can damage the spinal cord and/or optic nerves during attacks. It is a demyelinating condition, meaning, it damages the protective myelin sheath around the nerve fibres. In many ways, NMO can be confused with multiple sclerosis (MS) however NMO is less common than MS and attacks can be more severe than MS attacks. The cause of NMO is unknown.

Who gets NMO?

NMO is most commonly seen in women between the ages of 20 and 40. However, NMO has been diagnosed in children as young as 2 years old and adults in their 60's.

What is the difference between NMO and MS?

- Attacks of NMO are usually more severe with lasting disability.
- NMO and MS are different diseases and require different treatments.
- People with NMO have a detectable antibody that targets Aquaporin-4 (AQP4).

What are the symptoms?

Although the symptoms of NMO vary from person to person, they usually fall into two distinct types:

Inflammation of the optic nerve (optic neuritis)
This can cause blurring, or reduced or loss of vision. It typically affects only one eye, but can affect both. It can be painful, particularly when you move your eye. You may find your colour vision is affected too, with colours appearing 'washed out' or less vivid than usual. Symptoms are usually temporary, but they can become permanent. Optic neuritis tends to be more severe in NMO than in MS.

Inflammation of the spinal cord (transverse myelitis)
This can cause a range of symptoms, including pain in your neck or back; altered sensations such as numbness, tingling, coldness, or burning below the affected area of your spinal cord; weakness in your arms or legs; bladder and bowel problems; and muscle spasms. Transverse myelitis attacks also tend to be more severe in NMO than in MS.

How is NMO diagnosed?

A neurologist who specializes in NMO and MS is often required. Tests frequently include magnetic resonance imaging (MRI) of the brain and spinal cord, vision tests, and blood tests.

MRI (magnetic resonance imaging)
MRI is one of the main tools in the diagnosis of NMO. It will show where there has been inflammation in your brain or spinal cord – these appear as white areas as seen on MRI.

Generally, if you have NMO, the MRI of your spine will often show inflammation over three or more segments of your spinal cord – and the lesions will usually be longer than would be the case with MS.[2] An MRI of your brain may appear normal with NMO – although sometimes it can show signs of some inflammation similar to MS.

Lumbar puncture
A lumbar puncture is used to collect a small amount of cerebrospinal fluid (CSF), the clear liquid which surrounds your brain and spinal cord. If this is done during an acute attack of transverse myelitis, there may be increased white blood cells and raised proteins. This test will check for a particular antibody, known as **oligoclonal bands** which is usually found in the CSF of people with MS and less commonly in NMO.

Evoked potentials
This involves testing the time it takes for your brain to receive messages. Your neurologist will place small electrodes on your head to monitor your brain waves responding to what you see or feel. This is a painless procedure. If myelin damage has occurred, messages to and from your brain will be slower.[1]

Blood test
Unlike MS, there is a blood test that can be carried out for NMO. The test checks for an antibody known as **aquaporin-4 antibody**. Although this antibody is specific to NMO, it's not a conclusive test – around 60% of people with NMO have it, so you may still have NMO even if you don't have the antibody.

How is NMO treated?

Although the symptoms of NMO and MS seem similar, they are different diseases with different pathologic mechanisms, therefore are treated in different ways. Disease-modifying drugs used to manage MS, such as beta interferon and glatiramer acetate, would not usually be prescribed for NMO. Studies looking at the effect of beta interferon on NMO have found, at best, that it is of no benefit, and, at worst, that it can make NMO worse.[4, 5]

There are four aspects to treatment in NMO:
- Treating the attack
- Preventing relapses
- Treating the residual symptoms of the relapse
- Rehabilitation

Treatment of NMO attacks

Standard treatment involves intravenous steroids, and sometimes, additional treatments to remove antibodies (intravenous immunoglobulin or plasmapheresis/plasma exchange). These treatments should be started as soon as possible at onset of a severe attack.

Steroids
A course of methylprednisolone, a type of corticosteroid, is usually given to manage acute attacks of NMO – either intravenously or orally. Steroids work to reduce inflammation and can decrease the severity and duration of NMO relapses.

Plasma exchange
If a course of corticosteroids hasn't helped your attack, or if your attacks have progressed, you may be offered plasma exchange (PE). This procedure involves taking some of your blood and mechanically separating the blood cells from the fluid (plasma). The blood cells are then mixed with a replacement solution, typically albumin or a synthetic fluid with properties like plasma. The solution with the blood is then returned to your body.[3] PE temporarily clears the blood of antibodies that attack the myelin.

Chemotherapy

If a course of PE doesn't help the attack, some doctors may use chemotherapy to 'reboot' the immune system. It is important to speak with your doctor about the range of possible side-effects from chemotherapy medications.

Prevention of NMO attacks

NMO is a treatable disease and prevention of attacks using common medications is recommended after careful assessment by a specialist. Regular moderate exercise is also helpful.

Long-term treatment

There is currently no cure for NMO. However, you may be prescribed an immunosuppressant – which slows down the activity of the immune system – to prevent further attacks. Symptoms such as neuropathy, pain, stiffness, muscle spasms, bladder and bowel control problems can be managed with various medications and therapies. NMO does not progress between relapses so preventing attacks is essential.

Other resources

The Neuromyelitis Optica (NMO) Clinic and Research Program (University of British Columbia Hospital)
The NMO Clinical and Research Program at UBC is focused on the cause and the treatment of NMO and is Canada's only NMO program that is focused on clinical service with a research component.
Web: nmo.vchri.ca

The Guthy Jackson Foundation (United States)
This charitable foundation funds research into NMO. Also has an online community for people to connect to others with NMO, or working within the NMO field.
Web: guthyjacksonfoundation.org

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[1] Wingerchuk, D. M. et al. (2007) The spectrum of neuromyelitis optica. *Lancet Neurology*, 6, 805-15.
[2] Weinshenker, B. G. (2007) Neuromyelitis optica is distinct from multiple sclerosis. *Arch Neurology*, 64, 901-3.
[3] Wingerchuk, D. M. (2007) Diagnosis and treatment of neuromyelitis optica. *Neurologist*, 13, 2-11.
[4] Sellner, J. et al. (2010) EFNS guidelines on diagnosis and management of neuromyelitis optica. *European Journal of Neurology*, 17, 1019-1032.
[5] Palace J. et al. (2010) Interferon beta treatment in neuromyelitis optica increase in relapses and aquaporin 4 antibody titers. *Arch Neurology*, 67, 1016-1017.

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