



Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Neuromyelitis optica spectrum disorder

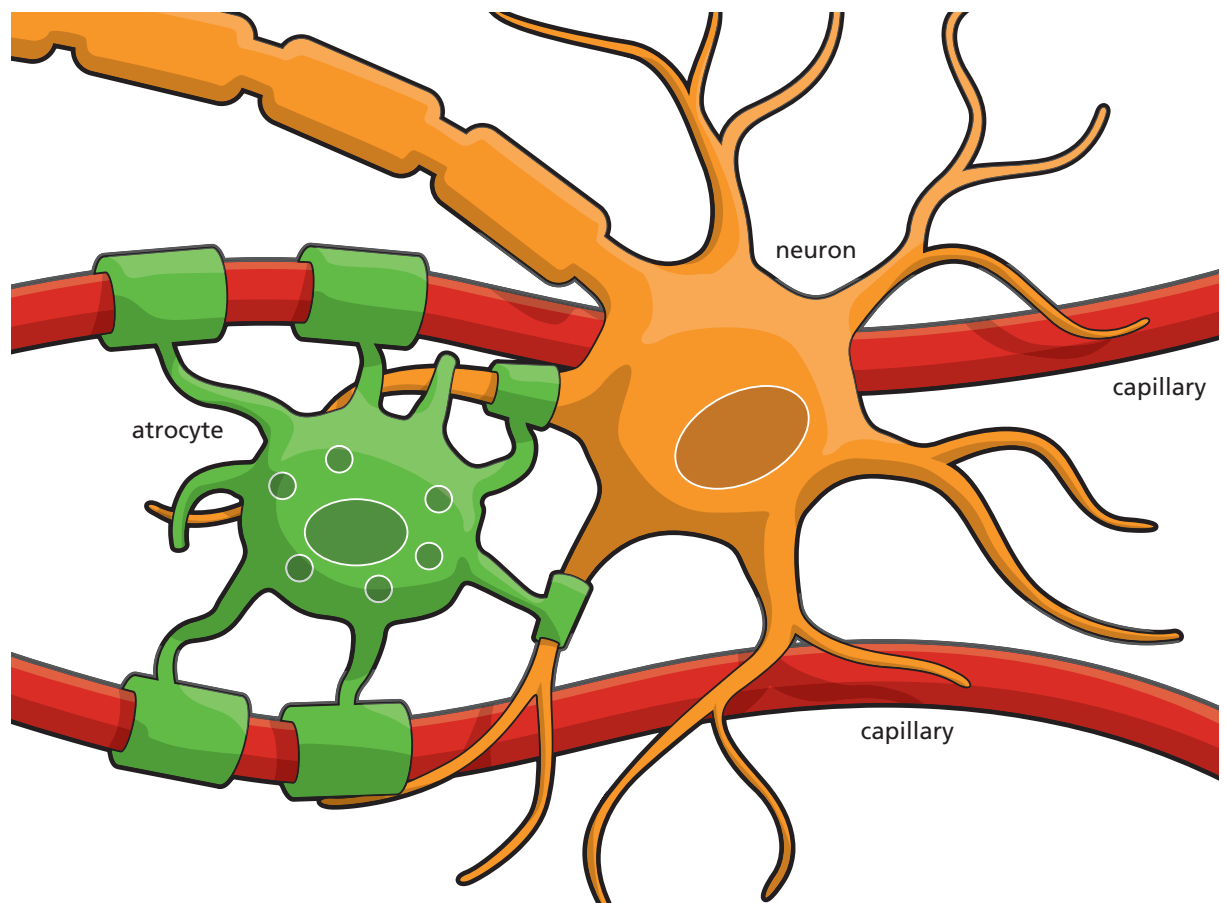
This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of neuromyelitis optica spectrum disorder (NMOSD) and where to get help.

What is neuromyelitis optica (NMO)?

Neuromyelitis optica spectrum disorder (NMOSD) is a rare neurological (brain) condition characterised by episodes of optic neuritis (inflammation or swelling of the optic nerve), transverse myelitis (inflammation or swelling of the spinal cord), together with one or more other diagnostic criteria including in some cases the presence of a specific antibody (AQP4). The episodes of inflammation are caused by the body's immune system becoming mis-

programmed and activating immune cells to attack the healthy specialised nerve cells in the optic nerve and spinal cord called astrocytes. Astrocytes are responsible for passing a variety of energy-giving substances from the blood to the nerve cells.

NMOSD is similar to multiple sclerosis in that it can relapse (as the nerves are attacked) and remit (improve) but the course and treatment of the condition is different.





What causes neuromyelitis optica spectrum disorder?

Neuromyelitis optica spectrum disorder is thought to be an autoimmune disorder although further research is needed to prove the actual mechanism of how the condition develops. Autoimmune disorders occur when the body mistakenly attacks itself rather than a foreign invader such as a bacterium or virus. We do not know exactly why this happens, sometimes it can follow on from a viral infection, but it can also occur with no prior warning or trigger event.

Research has shown that around 60 to 70 per cent of young people diagnosed with neuromyelitis optica spectrum disorder have a specific antibody in their blood. This antibody is known as 'aquaporin 4' or AQP4. The discovery of this antibody often confirms the diagnosis and helps rule out other demyelinating disorders.

What are the signs and symptoms of neuromyelitis optica spectrum disorder?

Symptoms are variable from person to person and can range from mild to severe depending on which parts of the body are affected. Neuromyelitis optica spectrum disorder normally involves episodes of optic neuritis as well as transverse myelitis. It can also present with episodes of nausea, vomiting and dizziness.

Optic neuritis can affect one or both eyes and the symptoms can include pain on moving the eye, blurred vision, loss of colour vision or even complete loss of vision. The symptoms of acute transverse myelitis can come on quickly and depend on where in the spine the swelling occurs. If the swelling is in the lower back, then symptoms can include back pain, weakness of both legs and bowel and bladder problems. If the attack is higher such as around the neck (cervical) area, then both arm and leg weakness can occur, and in severe cases breathing can be affected. As well as optic neuritis and transverse myelitis, the brainstem (known specifically as the area postrema) can also become inflamed leading to nausea, vomiting and hiccups.

How is neuromyelitis optica spectrum disorder diagnosed?

Neuromyelitis optica spectrum disorder can be mistaken for other similar appearing conditions, so diagnosis at a specialist centre is advised.

Neuromyelitis optica spectrum disorder tends to develop in adults and is rare in young people. Children developing the condition are usually diagnosed between the age of 10 and 14 years old, although it can develop at any stage of childhood.

Imaging scans such as MRI are used to identify the brain and spine changes. A lumbar puncture may also be used to look at a sample of the cerebrospinal fluid that surrounds the brain and spinal cord. Specialised eye tests called visual evoked potential (VEP) and electroretinogram (ERG) will also be used to look at the optic nerve and whether it is affected. Now that the antibody involved in neuromyelitis optica spectrum disorder has been identified, a blood sample can be examined in the laboratory to see if it is present. However, around 30 to 40 per cent of children and young people with neuromyelitis optica spectrum disorder do not have the antibody.

How is neuromyelitis optica spectrum disorder treated?

Steroid treatment can reduce some symptoms and stop new symptoms from developing. When the condition is first apparent, steroids are usually given into a vein once a day for three to five days. If there is no sign of improvement within a few days, other treatments may need to be considered. Intravenous immunoglobulin can be given, as can a treatment known as plasma exchange. This would be discussed in full detail if needed.

Once symptoms have improved, children often need to have maintenance treatment to prevent further episodes. Medicines called azathioprine, mycophenylate mofetil and sometimes rituximab, with or without steroids, are used to damp down the immune system.



What happens next?

Each episode can cause lasting damage to the nerves disrupting transmission of messages between the brain and body. Some children are left with lasting effects such as visual impairment, weakness in bowel and bladder function and movement problems. Preventive maintenance treatment aims to decrease the risk of disability.

Further information and support

NMO-UK is part of the national NMO specialist service and can offer information and support. Visit their website at www.nmo-ukresearchfoundation.org

The **Transverse Myelitis Society** offers information and support to anyone affected by neuromyelitis optica. Call them on 020 8568 0350 or visit their website at www.myelitis.org.uk

The **MS Society** supports people affected by multiple sclerosis and other demyelinating disorders. Call their helpline on 0808 800 8000 or visit their website at www.mssociety.org.uk and www.youngms.org.uk

The **British Brain and Spine Foundation** helpline is run by neurological nurses. Call them on 0808 808 1000 or visit their website at www.brainandspine.org.uk

The **British Trust for the Myelin Project** supports researchers and other people with an interest in demyelinating disorders. You can call them on 0161 292 3191 or visit their website at www.myelinproject.co.uk

The **Guthy-Jackson Charitable Foundation** in the United States supports people with Neuromyelitis Optica. Visit their website at www.guthyjacksonfoundation.org