Neuromyelitis Optica Spectrum Disorder (NMOSD) is a neurological **autoimmune condition** characterised by inflammation of the optic nerves and/or spinal cord. For some people the brain or brainstem may be affected.

What is an autoimmune condition?

A disease where the body's own immune system attacks healthy parts of its own organs and cells.

Antibodies:

Some patients have certain proteins (antibodies) in their blood which cause the disease and when detected by a blood test, confirm the diagnosis. These can be:

- **Aquaporin 4 (AQP4) antibody**
- **Myelin Oligodendrocyte Glycoprotein (MOG) antibody**

Antibody negative NMOSD

Some patients have the symptoms of NMOSD but have no antibodies. It is possible they have an as yet undiscovered antibody.

How is NMOSD diagnosed?

1. An episode of one of the following clinical events:
   - Optic neuritis (which is usually more severe than standard optic neuritis or both eyes are affected at once, see below for symptoms)
   - Transverse myelitis (usually a long spinal cord lesion the length of at least three vertebrae, see below for symptoms)
   - Area postrema syndrome (unexplained and persistent hiccups or nausea and vomiting)
   - Acute brainstem syndrome (vertigo, dizziness and severe imbalance)
   - Other parts of the brain may be affected but this is very rare
2. Positive test for AQP4 antibody
3. MRI abnormalities expected with NMOSD
4. Exclusion of alternative diagnoses

Key features of NMOSD

AQP4 antibody NMOSD:

- In the UK there are about 700 people with AQP4 antibody NMOSD
- It may be more common in those of Asian and African descent
- Predominantly affects women, with up to 5 women being affected for every man

All types of NMOSD:

- Can affect people at any age, including children (more rarely) and the elderly
- NMOSD is not hereditary
- NMOSD is not infectious

What may happen in NMOSD?

Optic Neuritis:

'Inflammation of the optic nerve'

Symptoms –

- Pain behind the eye particularly when it is moved
- Blurred vision
- Problems seeing colours
- Visual field defect; part of the vision is disturbed
- complete visual loss

Transverse Myelitis:

'Inflammation of the spinal cord'

Symptoms –

- Muscle weakness in legs and/or arms
- Altered sensations (pins and needles, numbness, tight band around chest or abdomen)
- Bladder and bowel problems
- Severe pain

Brain or Brainstem:

'Inflammation of the brain or brainstem'

Symptoms –

- Prolonged hiccups
- Persistent vomiting and nausea
- Excessive sleepiness during the day
- Limb weakness that is not related to a spinal cord attack

How do I recognise an attack/relapse and what should I do?

If you have any **new** symptoms or **worsening** of your current symptoms you may or may not be having an attack and it is not easy to know for sure by telephone. Therefore, if this happens, it is important to arrange an assessment with your GP and/or local neurologist.

As infections, such as bladder infections, can cause symptoms that are sometimes similar to an attack, we may ask you to have a urine sample checked and/or blood tests at your GP surgery.

We also ask you to inform your NMO Specialist Nurse so that we can update your records and advise your local team if they confirm a relapse or need advice.

The NMO nurses are available on the numbers below Mon-Fri 9-4pm: **Liverpool: 0151 529 8357 Oxford: 01865 231905.**

The medical team will ask you about your symptoms, your medications and other illness (such as colds, infections) so it is good to have this information to hand.

Medication to treat an attack

Urgent intravenous Methylprednisolone (IVMP) 1 gram per day for 3-5 days (oral dose is 500mg of methylprednisolone)

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If the attack is severe or does not respond to IVMP

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Plasma exchange for 5-7 days or intravenous Immunoglobulin (IVIG) or more IVMP

Medication to prevent attacks

If you have AQP4 antibodies you **will** need to take long-term immunosuppression. If you don't have AQP4 antibodies but have had recurrent attacks you **may** need to take long-term immunosuppression. This usually includes:

- Prednisolone 5-15mg per day **and** either

1. Azathioprine 2.5mg/kg per day or
2. Mycophenolate 2-3 grams per day or
3. Methotrexate 12.5-25mg per week

If you have an attack whilst taking an adequate dose of the above medications for at least six months your medication will need to be reviewed with the NMO team.

Rituximab infusions may then be tried.

You will always need to have regular blood tests at your GP whilst taking these medications

We recommend patients with AQP4 positive antibodies should not reduce immunosuppressant medications (e.g. prednisolone, azathioprine, mycophenolate, and methotrexate) without discussion with the NMO Team, or your Consultant Neurologist, due to the risk of relapse, unless a medical emergency requires this.

Recovery from a relapse and on-going symptoms

The time taken to recover after a relapse can vary hugely from person to person.

Over time many of your symptoms may improve of their own accord, aided by treatment received.

However, if the nerves have been damaged or destroyed and new pathways cannot be made, persistent symptoms or deficits may occur.

The range and severity of symptoms will be different for each individual.

Having a diagnosis of NMO or living with disability can also affect your psychological well-being.

On-going symptoms may include:

- reduced vision or blindness
- pain and spasms
- bladder, bowel and sexual problems
- muscle weakness or stiffness
- fatigue

If you are affected by any of these symptoms or worried about your NMO condition please speak to your NMO nurse in clinic or by telephone

For more information see **www.nmouk.nhs.uk**

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