What is Neuromyelitis Optica (NMO)?

Neuromyelitis Optica is due to inflammation (swelling) in the optic nerve (optic neuritis) and spinal cord (myelitis). It is a type of a "demyelinating" disorder. Myelin is the protective covering of the nerve fibres. In Neuromyelitis Optica the myelin that surrounds the nerves in spinal cord and optic nerve is damaged.

The optic nerve sends information from the eye to the brain about what is seen. The spinal cord transmits information between the brain and the legs, arms, bladder and bowel. Demyelination of spinal cord and optic nerve slows or interrupts the messages being sent from the nerves, resulting in various symptoms.

What are the symptoms?

The symptoms are of optic neuritis and transverse myelitis.

The symptoms of Optic Neuritis can affect one or both eyes and can include:

- Blurred vision
- Loss of colour vision
- Complete loss of vision
- Eye pain

The symptoms of Transverse Myelitis can include:

- Back pain
- Weakness of both legs and sometimes arms
- Numbness or tingling in legs and sometimes arms
- Loss of bowl and bladder function- constipation and difficulty passing urine

The symptoms vary. Some children will have one attack of optic neuritis and transverse myelitis. Other children will have a number of attacks throughout life.

Generally only the optic nerve and spinal cord are involved. However in rare cases it can also affect parts of the brain and causes symptoms such as vomiting and hiccups.

What causes **Neuromyelitis Optica?**

- Neuromyelitis Optica is an "autoimmune" condition. It often follows an infection such as a cold. The immune system protects the body from things like bacteria and viruses. Autoimmune diseases confuse the body's immune system. As well as fighting against the bacteria or virus, it attacks healthy cells and tissue. In Neuromyelitis Optica the immune system reacts against the myelin in the optic nerve and spine. The immune cells attack and damage the myelin coating of the nerve fibres. This slows or interrupts the messages being sent from the nerves.
- Sometimes a specific trigger cannot be identified.
- 70% of people with Neuromyelitis Optica have an antibody called Neuromyelitis Optica Immunoglobin G (NMO IgG) present in their blood. It is thought that NMO IgG may damage the water channel (aquaporin 4) surrounding the cells of the spinal cord and optic nerve.
- Autoimmunity is not infectious.

How is the diagnosis made?

There are several tests that a doctor may do. A doctor needs to make sure that the diagnosis is Neuromyelitis Optica and not another disorder. Tests may include:

- a medical history (the doctor will ask many questions)
- A clinical examination
- Brain MRI scan to look for swelling in the optic nerve and to make sure there is no swelling in the brain
- Spine MRI scan to check for swelling in the spinal cord and rule out causes other than *transverse myelitis*
- A lumbar puncture (spinal tap) to make sure that there is not an infection in the spinal fluid, such as meningitis or encephalitis
- Blood tests to look for infectious triggers and for other conditions which can be confused with Neuromyelitis Optica. The test also checks for the presence of the NMO IgG antibody.
- Special eye tests like the electrical function of the optic nerve (called visual evoked potentials)

What is the treatment?

Medications are used to reduce the inflammation (swelling) in the optic nerve. The main medication that is used is called methylprednisolone (a corticosteroids) given by an infusion (drip) once a day for three to five days. This may be followed by prednisolone, a medication that can be swallowed.

Many children improve with high doses of methylprednisolone. If this medication does not work, there are other treatments that can be tried (plasmapheresis or intravenous immunoglobulin therapy).

If experiencing pain, a pain management plan will be developed.

Physiotherapy and occupational therapy will help improve strength, balance and function

If children have recurrent episodes of transverse myelitis and/or optic neuritis, they may be given a medication that may prevent the number and severity of these relapses. Azathioprine may be used to suppress the body's immune system. This stops the defence mechanisms from attacking the body. Other medications that may be tried include mycophenolate, rituximab and cyclophospamide. The doctor will discuss the benefits and side effects before starting these medications.

What are the side effects of methylprednisolone and prednisolone?

Most children tolerate these medications very well. Some children can develop moodiness or other behavioural changes. Steroids can also cause increases in blood pressure and blood sugar, which the doctors and nurses will check for and treat if necessary. Steroids can also irritate the stomach lining. A medication such as ranitidine will be given to prevent this stomach irritation. These medications may temporary suppress the immune system. The immune system may fight less well against viruses and bacteria.

What is the prognosis of Neuromyelitis Optica?

The outcome varies. Some children will have one attack of optic neuritis and transverse myelitis and make a near complete recovery. Other children will have a number of attacks throughout life and have permanent disability with reduced vision and inability to walk.

For each attack it is possible to make a nearly complete recovery or have residual symptoms such as blurred vision or decreased colour vision, weakness in both legs. For most children, recovery begins within days and continues for up to one year.

Will school performance be affected?

Because Neuromyelitis Optica affects the optic nerve and spinal cord (which are not involved in thinking), most children return to their original school performance. However, some children may have difficulty with their school work due to missed school days during the illness as well as changes in vision and handwriting. Mobility difficulties may make it difficult to get around school. Some children experience tiredness or fatigue and part-time schooling can be helpful. If there are any changes in the school work, it is important to let

the doctor know as well as the SENCO at school know, so that they can work together to develop an educational plan.

Will it happen again? Will symptoms occur with every infection?

In some patients, it can re-occur. It is not known why children develop Neuromyelitis Optica with certain infections at certain times. If new symptoms occur,, such as changes in vision and gait, it is important to let the doctor know immediately.

How is Neuromyelitis Optica similar to multiple sclerosis (MS)?

Both involve autoimmune responses to myelin. They are both "demyelinating" disorders. Symptoms common to both disorders include loss of vision and weakness. It can be difficult sometimes for doctors to tell the difference. However there are clues in the blood (NMO antibody) and on brain MRI to try to distinguish Neuromyelitis Optica from MS. Corticosteroids are used to treat attacks of both Neuromyelitis Optica and MS. Most patients with MS and Neuromyelitis Optica are treated with ongoing medication to prevent attacks. The medications used to prevent attacks can be different.