

# The Transverse Myelitis Association

...advocating for those with acute disseminated encephalomyelitis, neuromyelitis optica, optic neuritis and transverse myelitis



## OPTIC NEURITIS

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### OVERVIEW

Optic neuritis (ON) is an inflammatory demyelinating condition of the central nervous system that results in the loss of vision and is associated with eye pain, loss of color vision and visual field deficits. While ON can occur in isolation, it is often part of multiple sclerosis (MS), Acute Disseminated Encephalomyelitis (ADEM) or neuromyelitis optica (NMO or Devic's disease). ON can be the presenting feature of MS (15-20% of the time) and occurs in 50% percent of those diagnosed with MS at some point during their illness.<sup>2</sup> ON is typically monocular (affecting one eye), though it can also affect both eyes sequentially or simultaneously. Bilateral ON tends to be more common in children younger than 15 years old.<sup>2</sup>

The most common cause of ON is inflammatory demyelination of the optic nerve. The pathology (similar to that of acute MS) involves plaques in the brain, with perivascular cuffing, edema in the myelinated nerve sheaths, and myelin breakdown.<sup>2</sup> Similar to MS, a genetic susceptibility for ON is suspected, and it is believed that the demyelination in ON is immune-mediated. However, the specific mechanism and target antigen(s) are unknown.<sup>2</sup>

### EPIDEMIOLOGY

ON is a rare condition. US studies estimate the annual incidence to be 6.4 per 100,000 in population.<sup>2</sup> The occurrence of ON tends to be the highest in populations located in higher latitudes (in the northern US and Western Europe), and is the lowest in regions closer to the equator.<sup>2</sup> ON is more common in women (predominance ratio of 3:1), and develops in most patients between the ages of 20 and 45.<sup>3</sup> Additionally, ON typically occurs more frequently in Caucasians than African Americans.<sup>2</sup> It has been reported that whites with northern European descent develop ON eight times more frequently than blacks and Asians.<sup>3</sup>

### SIGNS AND SYMPTOMS

The classic symptoms of acute ON consist of unilateral loss of vision (in 70 percent of individuals), periocular pain,

and dyschromatopsia (color blindness or color vision deficiency).<sup>3</sup> This typically comes on over the course of a few days and peaks within one to two weeks.<sup>2</sup> ON usually begins with decreased vision in one eye. Approximately 90 percent of diagnosed individuals also experience pain behind the eye which is usually exacerbated by eye movement.<sup>3</sup> Visual loss can vary from mild reduction and minor blurring to no perception of light. Symptoms tend to exacerbate with increased body temperature.<sup>3</sup> Other common signs and symptoms of acute ON include: visual field defects, swelling of the optic nerve, photopsias (the presence of perceived flashes of light), and an afferent pupillary defect always occurs in ON if the other eye is uninvolved.<sup>2</sup>

Another key aspect of ON is that vision and eye pain usually improve within 2 to 3 weeks after the onset of symptoms. More than 90 percent of individuals experience visual improvement within this timeframe regardless of treatment.<sup>3</sup> Should symptoms persist for longer than 3 weeks, it suggests that it is either an atypical type of ON or is a different diagnosis.

## DIAGNOSIS

Generally, a clinical diagnosis of ON is based on the history and examination findings. Though demyelination is its most common identifiable cause, many other causes of optic neuropathy may resemble ON, and misdiagnosis is not uncommon.<sup>3</sup> Diagnostic testing is typically directed toward excluding other causes of visual loss in atypical cases, and assessing the risk of subsequent MS.<sup>2</sup> An early evaluation is essential to ensuring visual recovery has begun and to reconsider the diagnosis if it has not.

As mentioned, in typical cases of ON, visual improvement occurs within 2 to 3 weeks regardless of treatment. Thus, in typical cases, which show no additional clinical signs and symptoms of a systemic disease, the value of diagnostic testing is fairly low. However, if there are atypical signs and symptoms (i.e., bilateral presentation, younger than 15 years old, or possible infection) suggesting an alternative diagnosis, a complete assessment should be undertaken.<sup>3</sup>

MRI is used to take images of the brain and orbits to confirm the diagnosis of ON. However, the real value of MRI in typical ON is not to image the optic nerves, but rather to image the brain as a prognostic indicator for the future development of MS.<sup>3</sup> Often the brain MRI shows white matter abnormalities, or lesions, which are characteristic of MS - ovoid, periventricular, and larger than 3 mm lesions which indicate a higher risk of developing MS.<sup>2</sup>

Lumbar puncture is usually not considered an essential diagnostic test in ON, but should be considered in atypical cases. Approximately 60 to 80 percent of ON diagnoses show nonspecific abnormalities in cerebrospinal fluid (CSF).<sup>2</sup> Additionally, 56 to 69 percent of individuals also show oligoclonal bands (OCB) in their CSF, which implies a higher risk of developing MS.<sup>2</sup> However, since OCB is closely associated with white matter lesions seen in MRI, the presence of OCB is not of high prognostic importance.<sup>2</sup>

Finally, optical coherence tomography (OCT) is also commonly used to detect ON. OCT measures the thickness in the retinal nerve fiber layer and detects thinning in 85 percent of patients with ON.<sup>2</sup> While lower values correlate with impaired visual outcome, its usefulness as a prognostic tool is limited due to the fact that abnormal values do not show up until early swelling disappears.<sup>2</sup> OCT is also important as a number of studies have found that a greater severity of optic nerve injury, seen on OCT, suggests NMO rather than ON associated with multiple sclerosis.<sup>2</sup>

Additional diagnostic tests used to detect ON or assess the risk of other conditions include: fluorescein angiography, visual evoked response, and Aquaporin-4-specific serum autoantibody to rule out NMO.<sup>2</sup>

## ACUTE TREATMENT

Corticosteroids are the most common treatment for ON. They may be given intravenously or as high dose tablets. While corticosteroids have been effective in improving short-term visual recovery, they do not seem to affect the long-term outcome. Due to the lack of long-term benefit and the risk of potential side effects (including: insomnia, weight gain, and mood alterations), the use of corticosteroids is usually not advised.<sup>3</sup> However, there are specific situations where they may be used to reduce the period of impairment and are usually considered when a more rapid recovery is required (such as patients with severe bilateral visual loss or those with occupations that require normal visual acuity).<sup>3</sup>

Unfortunately, there is currently no treatment that can reverse vision loss caused by ON.

## PROGNOSIS AND MANAGEMENT

Most people recover well from ON. In approximately 80 percent of individuals, vision tends to recover by itself starting within 2–3 weeks from the onset of symptoms, usually stabilizing over months and continuing to improve for up to 1 year.<sup>3</sup> According to a large clinical trial (the Optic Neuritis Treatment Trial), 1 year after the initial ON attack, 93 percent those diagnosed with ON had a visual acuity of 20/40 and 69 percent had visual acuity of 20/20.<sup>3</sup>

Additionally, the severity of initial visual loss does seem to affect final visual outcome and the best predictor of visual recovery is the baseline acuity at the time of the attack.<sup>3</sup> On average, visual function is worse when ON is an early presentation of MS.<sup>3</sup>

Even though recurrences of ON can occur, the long-term outcome remains good. ON can recur either in the same or the contralateral eye. After ten years of follow-up in the previously mentioned Optic Neuritis Treatment Trial, 35 percent of participants experienced at least one documented recurrence.<sup>3</sup> Long-term follow-up studies have shown that only two percent are left with significant visual impairment in both eyes.<sup>1</sup> Not surprisingly, recurrence is more common in those who are later diagnosed with MS.<sup>1</sup>

In some cases, where there is no response to steroids (either intravenous or oral), plasma exchange is considered as a therapy. Long term immunomodulation and MS therapies (interferon beta-1a and interferon beta-1b) can be used to delay the progression or onset of MS in individuals who are likely to be diagnosed as MS.

## REFERENCES

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