

Optic nerve

Optic nerve

- The optic nerve is approximately 50 mm long
- It extends from the lamina cribrosa upto the optic chiasma
- It can be subdivided into four segments:
 - **Intraocular** segment (optic nerve head)
 - **Intraorbital** segment
 - **Intracanalicular** segment
 - **Intracranial** segment

Optic nerve

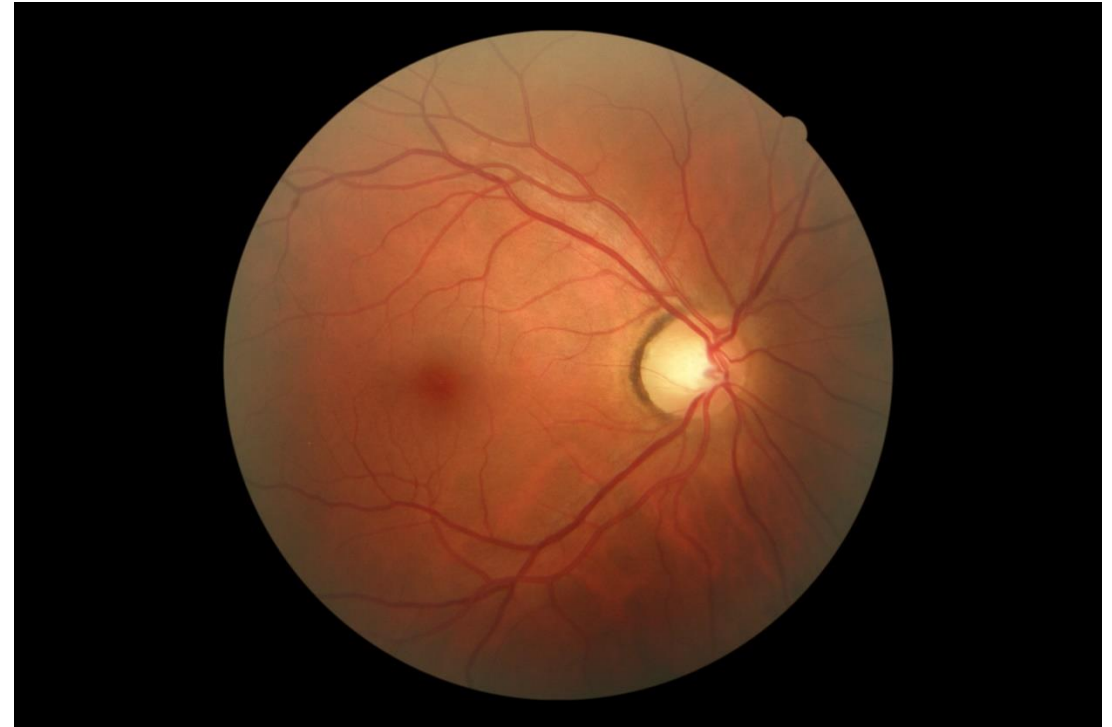
- The optic nerve carries approximately 1.2 – 1,5 million afferent nerve fibres, each of which originates in a retinal ganglion cell
- The optic nerve is covered with the meningeal sheaths, i.e. the pia mater, arachnoid mater and dura mater after it pierces the lamina cribrosa
- These meningeal spaces are continuous with those in the brain

NB

The optic nerve represents the most accessible part of the CNS, may be viewed directly using the ophthalmoscope and, now, the RNFL axons can be measured accurately using OCT

Intraocular segment (optic nerve head / optic disc)

- is the shortest, being 1 mm deep and approximately 1.5-2 mm in vertical diameter
- It has only nerve fibre layer so it does not excite any visual response “blind spot”
- It is a pink, oval or circular, sharp margins, there is a depression in its central part which is known as the “physiological cup” (the normal cup: disc ratio is 1:3 or 0.3.)



Intraorbital segment

- is 25–30 mm long and extends from the globe to the optic foramen at the orbital apex
- diameter is 3–4 mm because of the addition of the myelin sheaths to the nerve fibres
- At the orbital apex the nerve is surrounded by the tough fibrous annulus of Zinn, from which originate the four rectus muscles
- The intraocular and intraorbital parts are supplied by the branches of the ophthalmic artery, short posterior ciliary arteries and central retinal artery forming circle of Zinn

Intracanalicular segment

- traverses the optic canal and measures about 6 mm
- Unlike the intraorbital portion, it is fixed to the canal, since the dura mater fuses with the periosteum

Intracranial segment

- joins the chiasm and varies in length from 5 to 16 mm (average 10 mm).
- Long intracranial segments are particularly vulnerable to damage by adjacent lesions such as pituitary adenomas and aneurysms.
- The *intracanalicular and intracranial parts* are supplied by the branches of the anterior cerebral artery and ophthalmic artery.

Signs of optic nerve dysfunction

- **Reduced visual acuity** for distance and near is common
- **Relative afferent pupillary defect**
- **Dyschromatopsia** is impairment of colour vision, which in the context of optic nerve disease mainly affects red and green
- **Diminished light brightness sensitivity**
- **Diminished contrast sensitivity**
- **Visual field defects**, which vary with the underlying pathology

Classification of optic neuropathy by cause

- **Inflammatory** (Optic neuritis)
- **Glaucomatous**
- **Ischaemic** (Anterior non-arteritic, anterior arteritic, posterior ischaemic and diabetic papillopathy)
- **Hereditary** (Leber hereditary optic neuropathy, other hereditary optic neuropathies)
- **Nutritional and toxic**
- **Papilloedematous** (Secondary to raised intracranial pressure)
- **Traumatic**
- **Compressive** (Including secondary to an orbital lesion)
- **Infiltrative** (Inflammatory conditions (e.g. sarcoidosis), tumours and infective agents)



Optic neuritis

„ the eye as a window to the brain “

Definition and Classification

- *Optic neuritis is an inflammatory condition that affects the optic nerves*
- Acute optic neuritis is the most common optic neuropathy affecting young adults
- **Classification of ON according to clinical presentation:**
 - Typical optic neuritis is associated with multiple sclerosis
 - Atypical optic neuritis is manifestation of other diseases or conditions

- **Classification of ON According to ophthalmoscopic appearance:**

- Retrobulbar neuritis
- Papillitis
- Neuroretinitis
- Perineuritis

- **Classification of ON According to aetiology:**

- **Demyelinating.** The most common cause.
- **Parainfectious**, following a viral infection or immunization.
- **Infectious.** This may be sinus-related, or associated with conditions such as cat-scratch disease, syphilis, Lyme disease, cryptococcal meningitis and herpes zoster.
- **Non-infectious** causes include sarcoidosis and systemic autoimmune diseases such as systemic lupus erythematosus, polyarteritis nodosa and other vasculitides.

Typical optic neuritis

Epidemiology

- Studies from Sweden and Denmark have reported an annual incidence of 4-5 cases of new-onset optic neuritis per 100 000 persons
- Patients living in temperate climates seems to be more affected than it does other races
- Women are affected twice as often as men
- First manifestation is usually at age 20-40
- healthy young adults

Clinical features

- Discomfort or pain in or around the eye is present in over 90% and typically exacerbated by ocular movement;
- Unilateral vision loss is acute to sub-acute in onset progressing over a period of hours to days
- Visual acuity (VA) is usually 5/15–5/50, but may rarely be worse.
- Dyschromatopsia (change in color perception, red and green desaturation)
- Central scotoma, decreased retinal sensitivity
- RAPD (Marcus Gunn pupil)

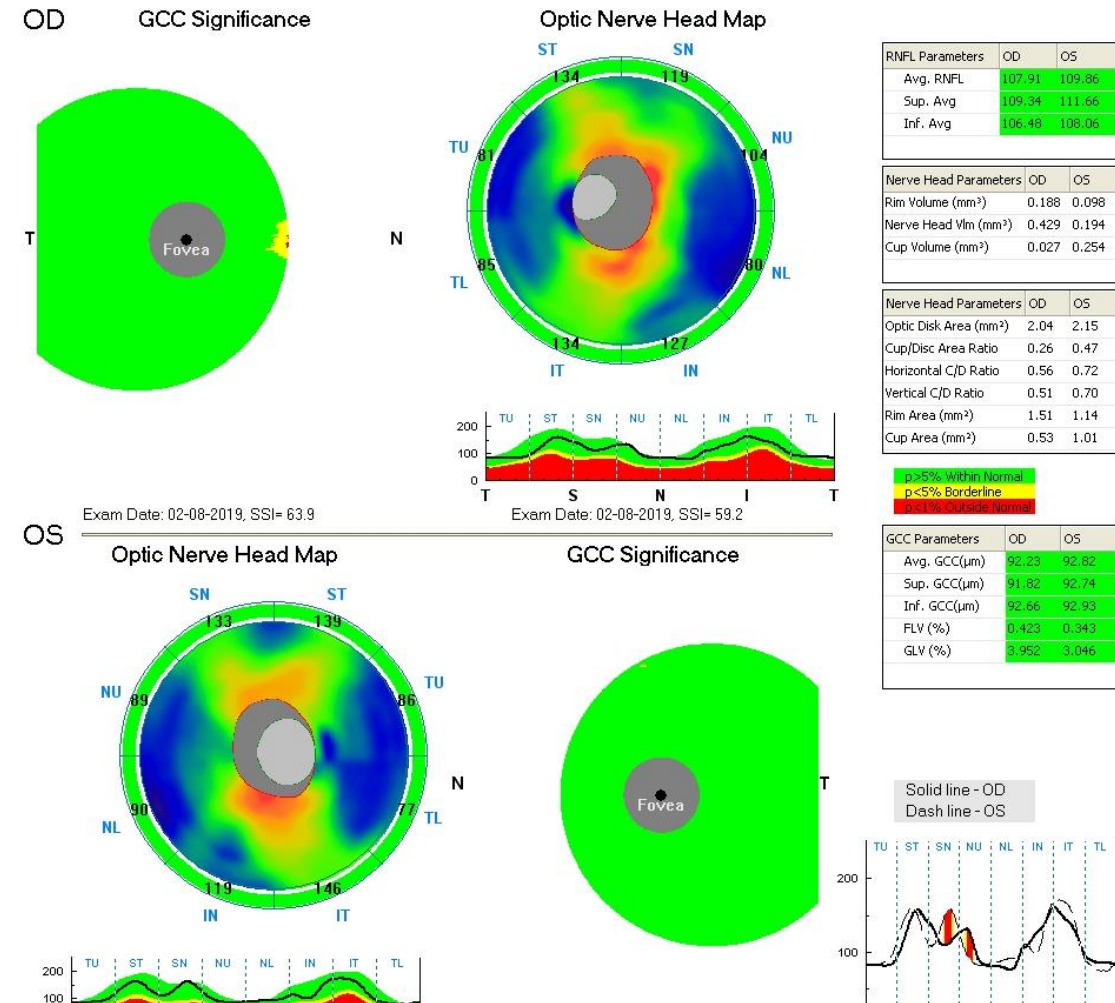


Ophthalmoscopy

- (65%) *retrobulbar ON* :optic disc appears normal
- is the most common type in adults
- (30%) *intraocular ON* : hyperaemia and oedema of the optic disc, peripapillary flame-shaped haemorrhages, cells in the posterior vitreous
- is the most common type of optic neuritis in children, but can also affect adults

Clinical features

- Patients with typical optic neuritis report improvement in pain within days, and vision begins to improve over the ensuing 2–4 week
- Optic nerve atrophy (loss of the unmyelinated axons in the retina) can readily be quantified by OCT
- RNFL thinning in MS-ON shows a definite predilection for the temporal quadrant
- Uhthoff phenomenon, in which vision loss is exacerbated by heat or exercise
- Pulfrich phenomenon, in which objects moving in a straight line appear to have a curved trajectory: Presumably caused by asymmetrical conduction between the optic nerves



Diagnosis

- History
 - Symptoms
 - Signs
 - VEP
 - MRI
 - OCT(atrophy)
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- Ancillary diagnostic tests: blood tests, serology, CSL exam., etc.

Treatment

Intravenous methylprednisolone (1 gm daily) for 3 days followed by oral prednisolone (1 mg/kg/day) for 11 days.

Vitamin B1, B6 and B12 injections are given in full doses.

Vitamin D

Atypical optic neuritis

Definition

- *Atypical optic neuritis is essentially optic neuritis from any other cause*
- **Atypical features include:**
 - male gender
 - age less than 18 years or greater than 50 years
 - bilaterality
 - absence of periocular pain
 - No light perception vision
 - vision loss that progresses past two weeks, and vision that does not improve for more than 3 months (either with steroids or spontaneously)
 - uveitis, severe disc swelling, atrophy, peripapillary hemorrhages macular exudates

Neuromyelitis optica (Devic's disease)

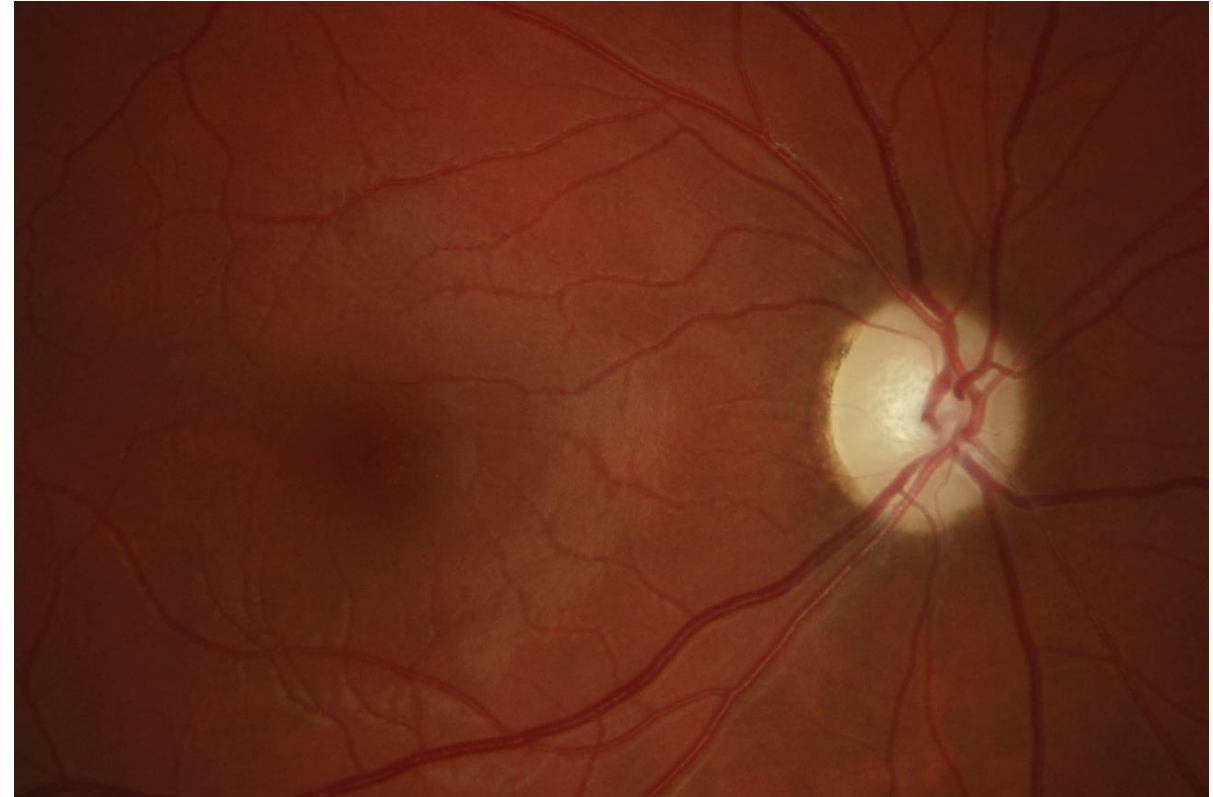
- *is a severe CNS inflammatory, demyelinating disease that preferentially affects the optic nerves and spinal cord*
- Is associated with antibodies against astrocyte aquaporin-4 water channels (AQP4-IgG) or MOG
- In a classic case of NMO, the patient has both optic neuritis and transverse myelitis extending over at least 2–3 segments of the spinal cord, with little or no abnormality in the brain

NMO-ON

- 1–3% of cases of optic neuritis
- Approximately 50% of NMO patients initially present with isolated ON, among whom 20% have bilateral involvement
- monophasic, relapsing-remitting
- female predilection
- the prevalence in women is up to 10 times higher than in men
- More commonly affect patients of Asian or African descent

NMO-ON

- is usually severe, typically bilateral or unilateral rapidly sequential to the fellow eye
- RAPD +/-
- vision loss is typically rapid and profound
- Abnormal color vision
- Field defects: altitudinal, arcuate
- **Ophthalmoscopy:**
- Retrobulbar ON
- papillitis (swollen disc) may be seen in one third of patients with optic neuritis



Parainfectious optic neuritis

- Association with viral infections such as measles, mumps, chickenpox, rubella, and may also occur following immunization.
- Children are affected much more frequently than adults.
- Presentation is usually 1–3 weeks after a viral infection, with acute severe visual loss generally involving both eyes.
- Bilateral papillitis is the rule
- The prognosis for spontaneous visual recovery is very good, and treatment is not required in the majority of patients.

Infectious optic neuritis

- **Sinus-related** optic neuritis is uncommon and is sometimes characterized by recurrent attacks of unilateral visual loss associated with severe headache and spheno-ethmoidal sinusitis.
- **Cat-scratch fever** (benign lymphoreticulosis) is usually caused by *Bartonella henselae* inoculated by a cat scratch
- **Syphilis** may cause acute papillitis during the primary or secondary stages



Infectious optic neuritis

- **Cryptococcal meningitis** in patients with acquired immunodeficiency syndrome (AIDS) may be associated with acute optic neuritis, which may be bilateral
- **Varicella zoster virus** may cause papillitis by spread from contiguous retinitis (i.e. acute retinal necrosis, progressive retinal necrosis)
- **Lyme disease** (borreliosis) is a spirochaetal infection caused by *Borrelia burgdorferi* transmitted by a tick bite It may cause acute retrobulbar neuritis, which may be associated with other neurological manifestations and can mimic MS.

Non-infectious optic neuritis

- Sarcoidosis
- affects 1–5% of patients with neurosarcoid
- It may occasionally be the presenting feature of sarcoidosis but usually develops during the course of established systemic disease.
- The response to steroid therapy is often rapid, and some patients require long-term low-dose therapy

- Autoimmune (SLE, vasculitis, ...)
- retrobulbar neuritis or anterior ischaemic optic neuropathy
- Some patients may also experience slowly progressive visual loss suggestive of compression
- Treatment is with systemic steroids and other immunosuppressants