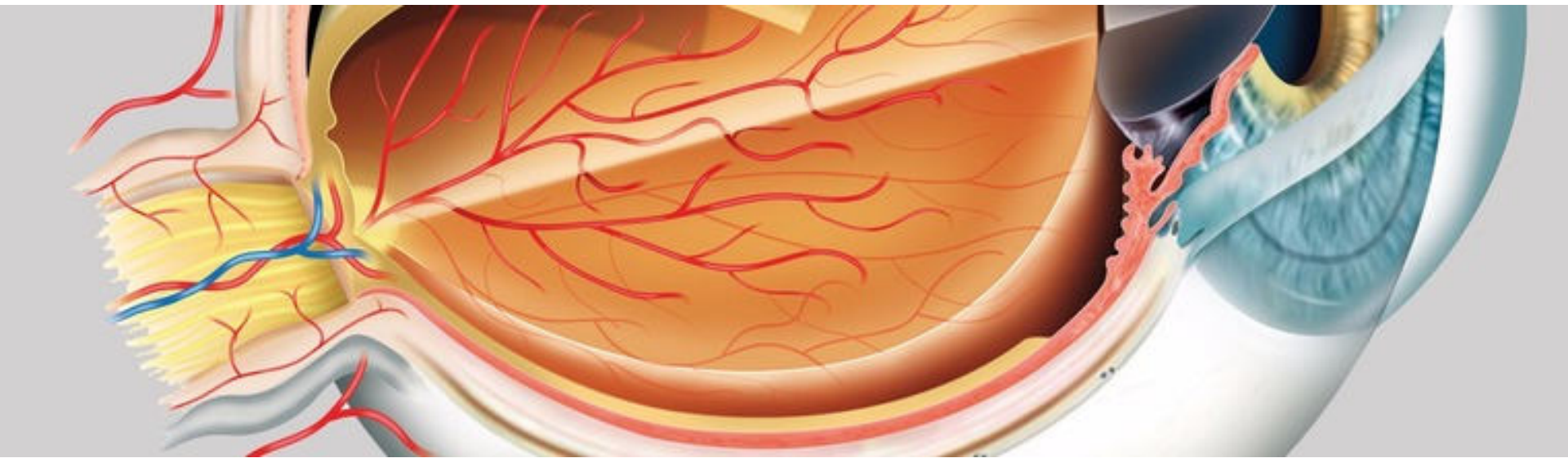
Optic Neuritis, Papilledema and Optic Atrophy



Department of Ophthalmology

Acknowledgement

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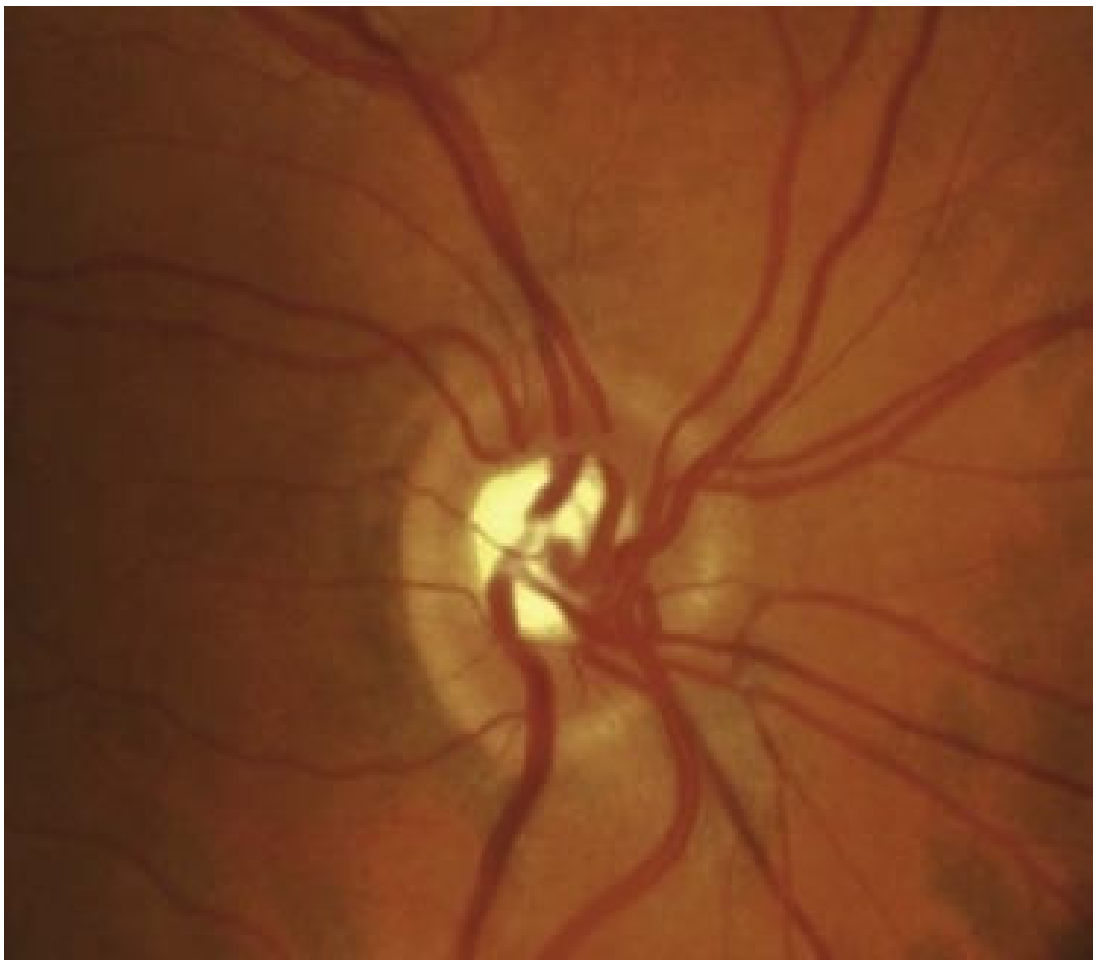
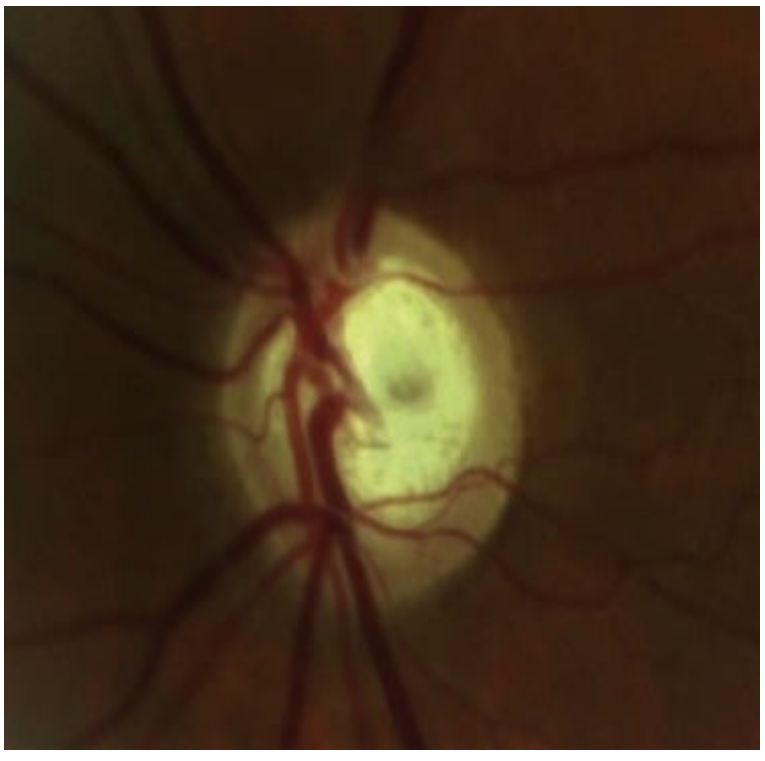
Learning Objectives

- At the end of the class, students shall be able to
- Define and classify optic neuritis.
- Understand the aetiology and principles of management of optic neuritis.
- Understand the stages and importance of papilloedema.
- Differentiate between various types of optic atrophy.

Normal optic disc



Question

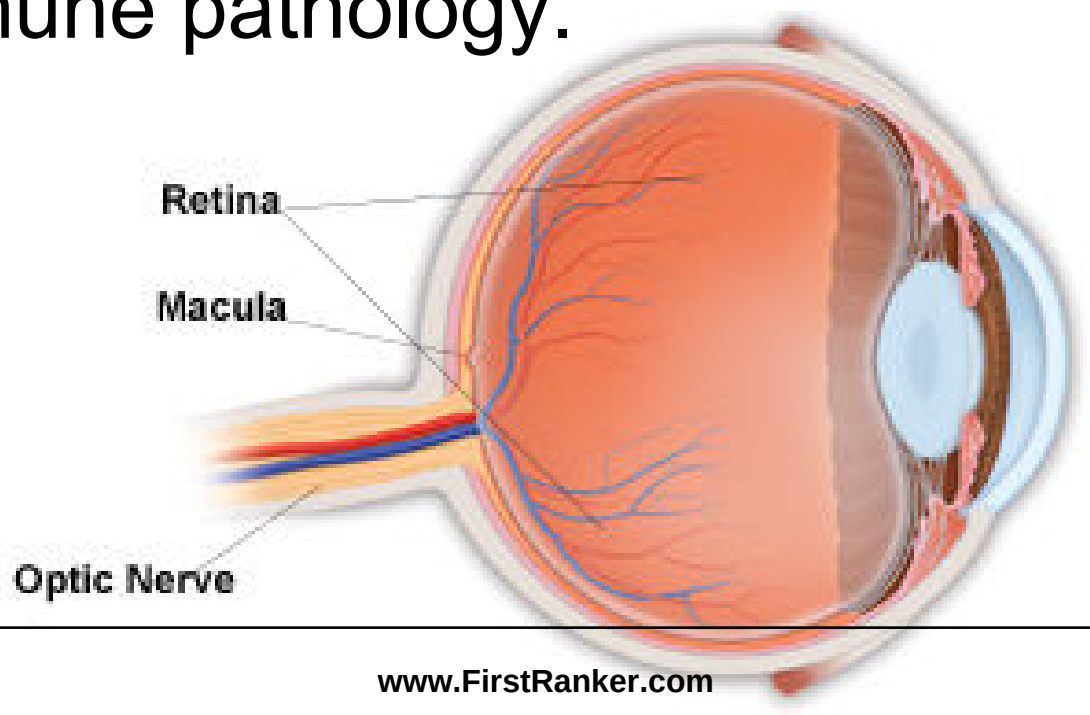


- The disc in question is of a 60 year old myope who is instilling timolol eye drops since the past 5 years.
- Is this a normal optic disc?

Optic Neuritis

Definition: Inflammation of the optic nerve, impairing nerve conduction.

Secondary to demyelination, infection or autoimmune pathology.



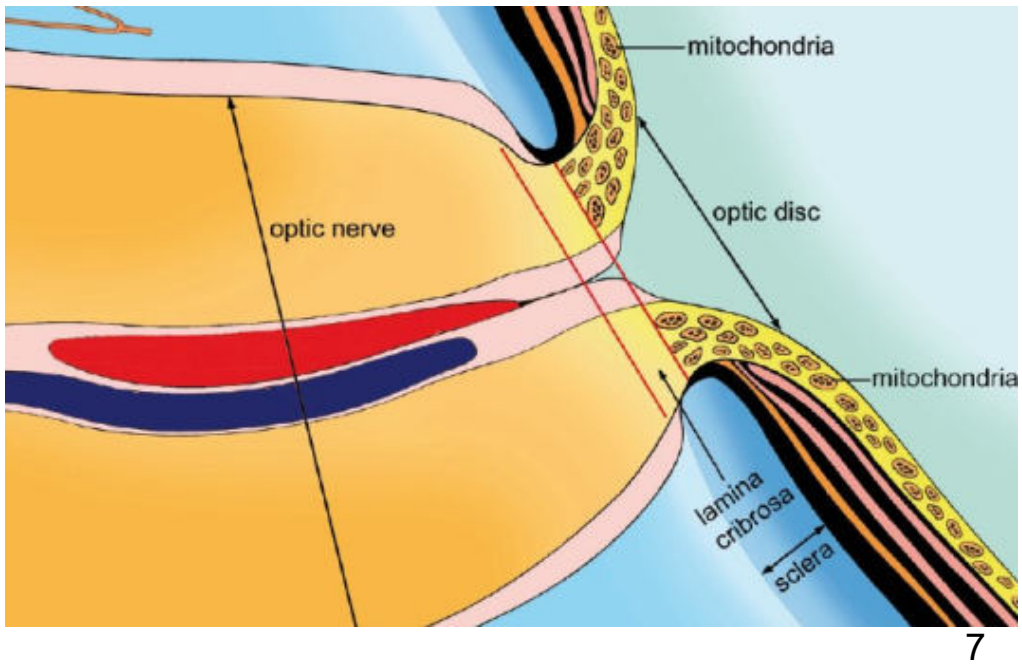
Classification

A. Papillitis

B. Retrobulbar neuritis
   - Acute
   - Chronic (toxic amblyopia)

C. Neuroretinitis

D. Perineuritis



Aetiology

- Idiopathic
- Demyelinating disorders

- Multiple Sclerosis
  - Presenting feature in 25% patients
  - 70% cases occur in established disease
  - Recurs in same/ opposite eye in 25% patients
  - **Uhthoff"s phenomenon:** impairment of vision more with increased body temperature
  - **Pulfrich phenomenon:** altered perception of moving objects

Aetiology

- Neuromyelitis optica (of Devic): acute, bilateral optic neuritis in young patient with paraplegia
- Post-viral: mumps, measles, chicken pox, whooping cough
- Metabolic/Nutritional deficiency:

  B1, B6, B12, B2, Folic acid deficiency

  Thyroid dysfunction, diabetes
- Hereditary optic neuritis (Leber's disease)

Aetiology

- Toxic amblyopia:

  Chloroquine, Ethambutol

  Tobacco, Ethyl alcohol, methyl alcohol

  Lead, Arsenic.
- Ischaemic: Giant cell arteritis, Takayasu's disease, PAN, SLE
- Granulomatous inflammation:

  Sarcoidosis, tuberculosis, syphilis

Symptoms

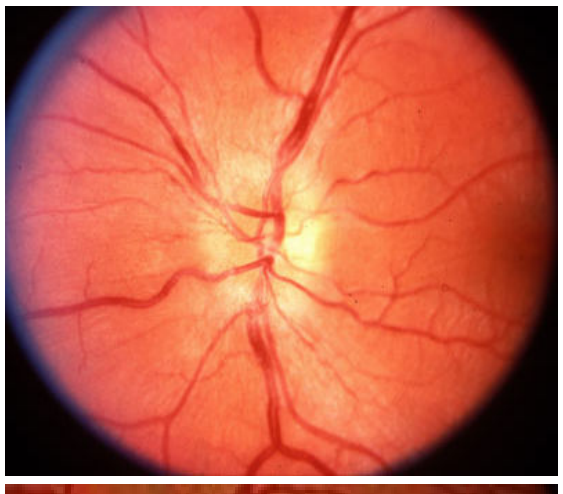
- Idiopathic/demyelinating : 20-40 years of age
- Viral: children
- Uniocular sudden/rapid diminution of vision
- Visual loss, usually maximum by end of second week, improves by 1-4 weeks
- Discomfort/pain behind eyeball especially when moved superiorly

Signs

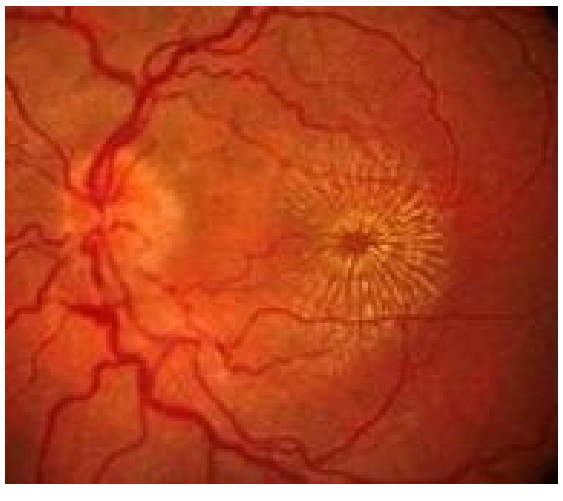
- **Visual Acuity:** Usually 6/60 or less
- **Local tenderness**
- **Pupillary reaction:** Sluggish, ill-sustained or RAPD
- **Impaired coloured vision:** hue, brightness
- Impaired contrast sensitivity
- Delayed dark adaptation
- **Visual Field:** central, centrocaecal or paracentral scotoma, more pronounced for coloured fields

Ophthalmoscopic findings

- **Optic neuritis:** MC in children, engorged, oedematous optic disc with obliteration of optic cup, small haemorrhages on disc



- **Retrobulbar neuritis:** MC in adults



- **Neuroretinitis:**

  Optic neuritis+ macular star

Differential diagnosis

- Papilloedema
- Pseudopapillitis

  High hypermetropia,

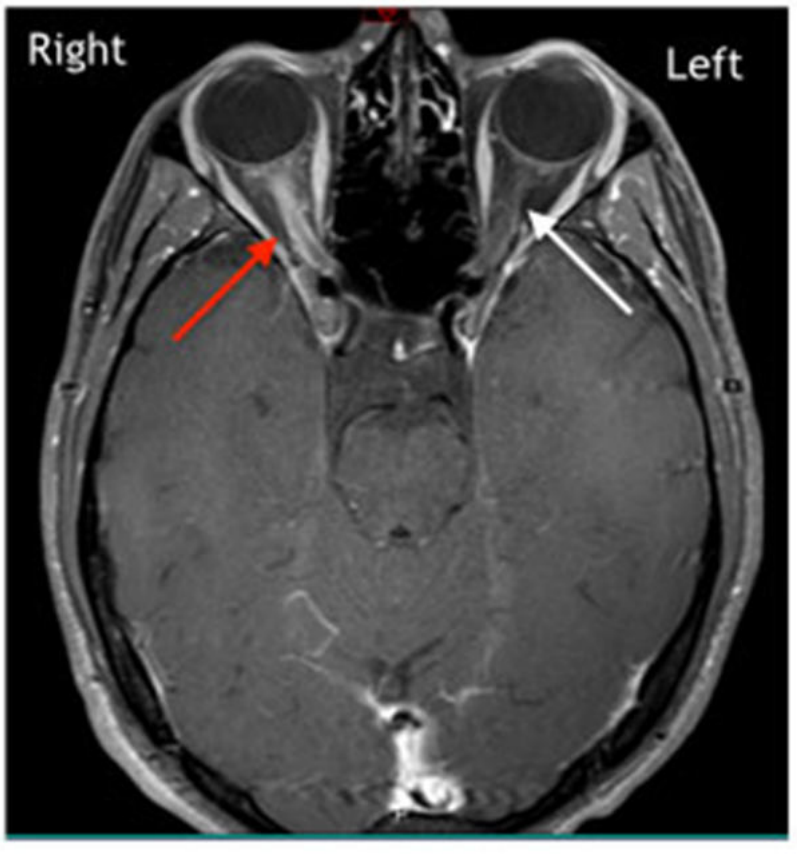
  Myelinated nerve fibres,

  Optic nerve head drusen

  (blurred margin, disc not significantly elevated, no vascular changes, stationary)

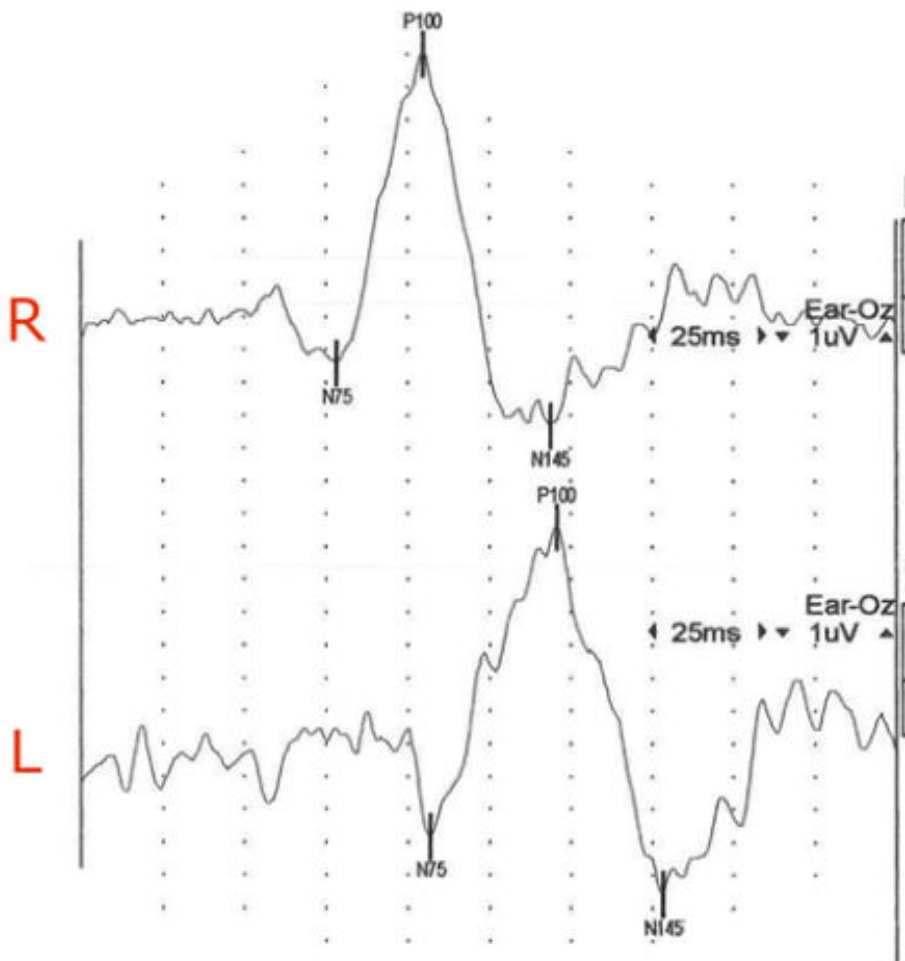
Investigations

- MRI: demyelinating lesions, SOL
- VEP: reduced amplitude and delayed transmission time (P100 latency increased)



Right optic neuritis

Early Optic Neuritis Nerve Asymmetry



RIGHT Eye VEP

Text	Lat N75 ms	Lat P100 ms	Lat N145 ms	PP Amp 75-100 uV
	1.1:N75	1.1:P100	1.1:N145	1.1:N75 P100
Oz	78.0	105	144	9.48

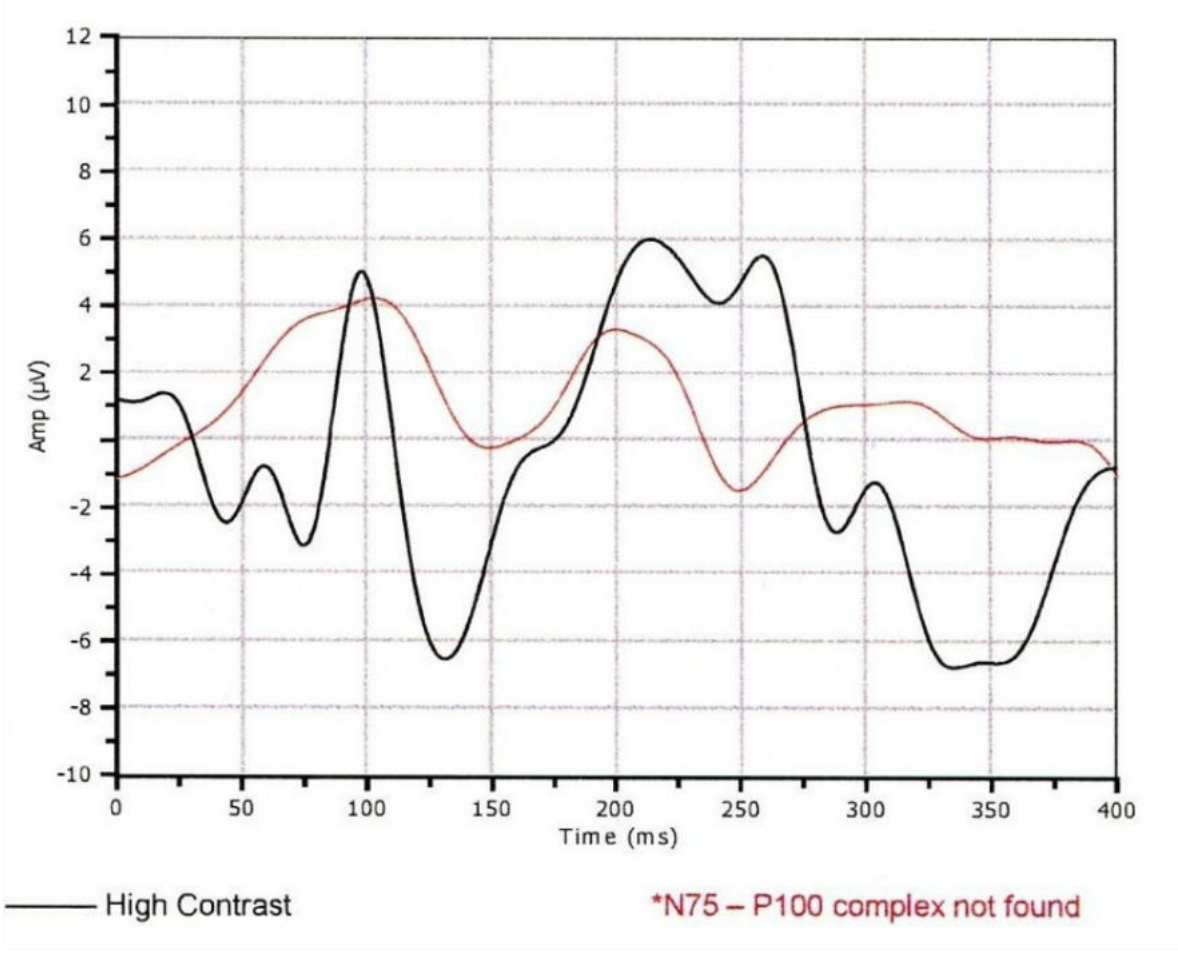
LEFT Eye VEP

Text	Lat N75 ms	Lat P100 ms	Lat N145 ms	PP Amp 75-100 uV
	2.1:N75	2.1:P100	2.1:N145	2.1:N75 P100
Oz	106	145	178	9.47

Interocular Comparison

Text	LatDiff LT-RT	PP Amp Ratio LT-RT
	40.8 msec	99.8 %

Advanced case of optic neuritis



Course and prognosis

- Recovery takes 4-6 weeks
- 90% recover normal VA, but colour vision defects may persist
- No correlation between initial visual loss and final visual outcome
- 10% secondary or post-neuritic optic atrophy
- Better outcome in young, unilateral cases

Treatment

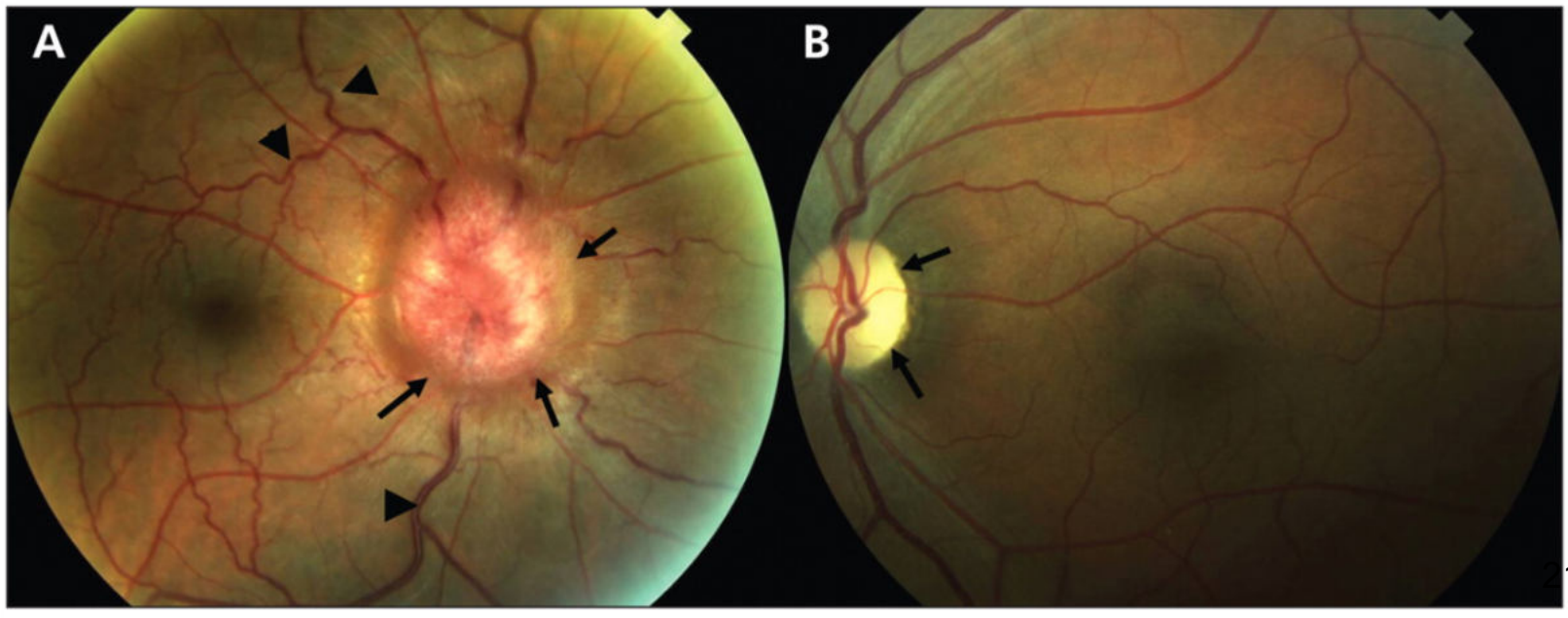
- Of cause e.g. anti-infective therapy
- Intravenous methyl prednisolone 20 mg/kg/day(250 mg QID) for 3 consecutive days followed by oral prednisolone 1-1.5 mg/kg
- Dexamethasone 200 mg OD pulse for 3-5 days is a cheaper alternative
- Supportive therapy

Papilloedema

- Definition: Bilateral, non-inflammatory passive swelling of optic disc due to raised Intracranial pressure.
- Does not develop if optic nerve is atrophic

Foster-Kennedy syndrome : contralateral papilloedema with ipsilateral pressure atrophy of optic nerve

Due to - frontal lobe tumour, olfactory meningioma



D/D: Causes of 'disc oedema'

- Papillitis, neuroretinitis
- Anterior Ischemic Optic Neuropathy
- Optic Nerve glioma, meningioma
- Central Retinal Venous Occlusion

Aetio-Pathogenesis

- Elevated Intracranial pressure due to any cause
- Prelaminar Optic Nerve is affected by changes in tissue pressure, IOP and CSF pressure
- Increased CSF pressure increases tissue pressure hampering axoplasmic flow
- This further increases pressure on pre-laminar capillaries and small veins causing vasodilatation and tortuosity
- Venous drainage compromise further increases congestion

General Symptoms

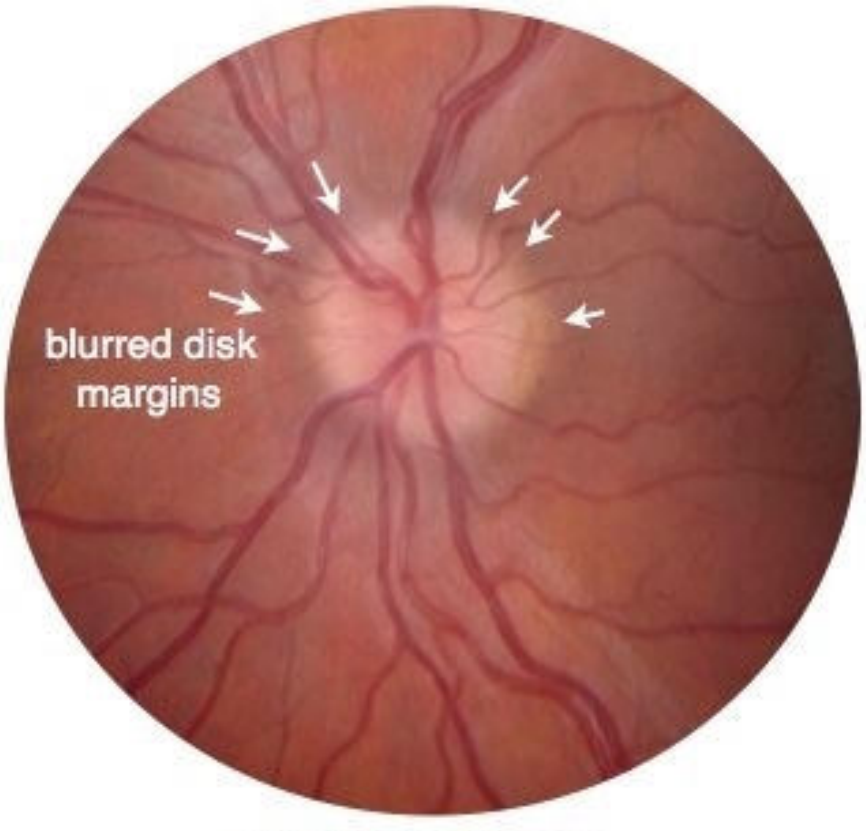
- Headache, made worse by coughing or straining
- Vomiting
- Focal neurological deficit with/without changes in level of consciousness

Ocular symptoms

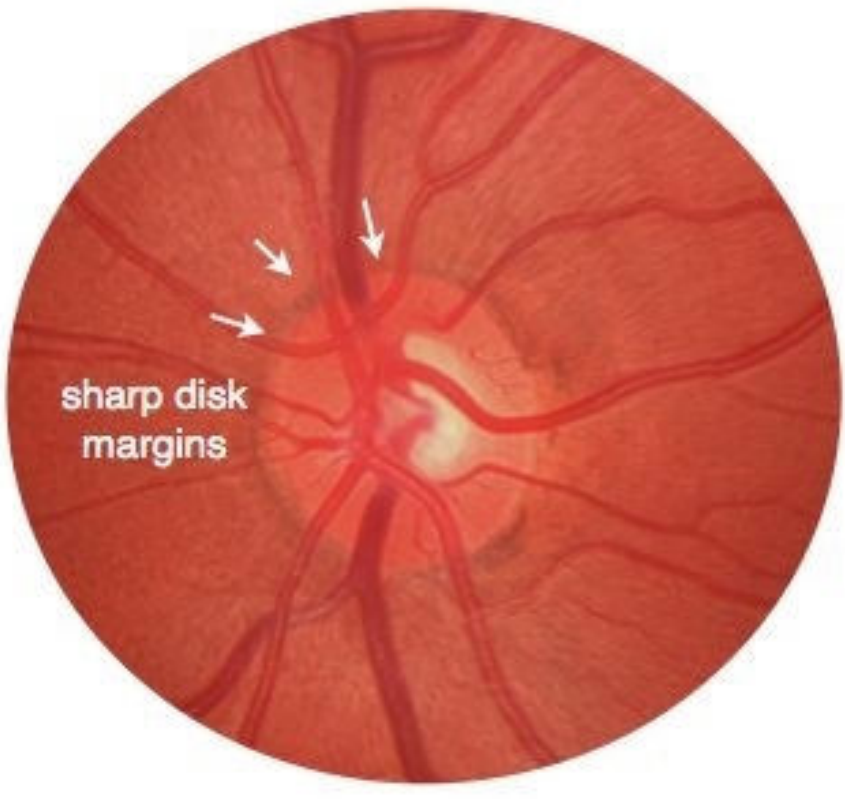
- VA may be normal until late stages
- Amaurosis fugax in some
- In 25% patients, visual symptoms occur only in severe, advanced papilloedema

Signs

- Pupillary reactions are normal until secondary atrophy sets in
- Early:
  - Blurring of nasal>superior>inferior margins of disc
  - Disc hyperemia and dilated capillaries
  - Spontaneous venous pulsation absent
  - Splinter haemorrhages at/just off disc margin
  - Normal optic cup preserved



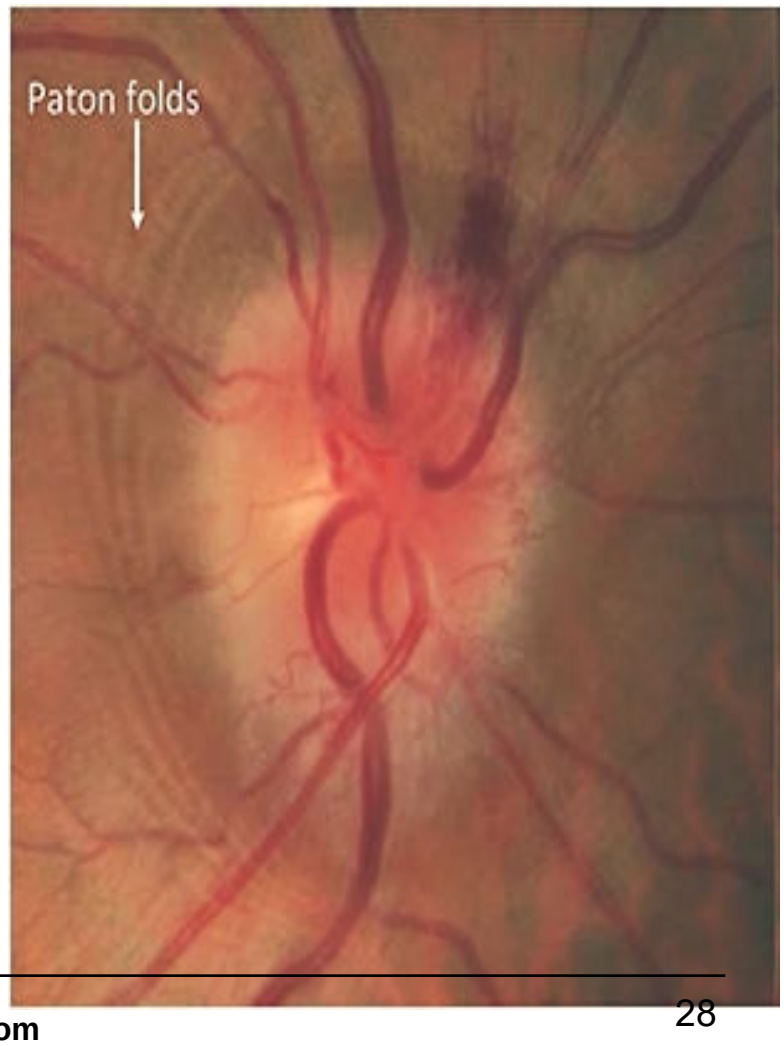
Papilledema



Normal Optic Disk

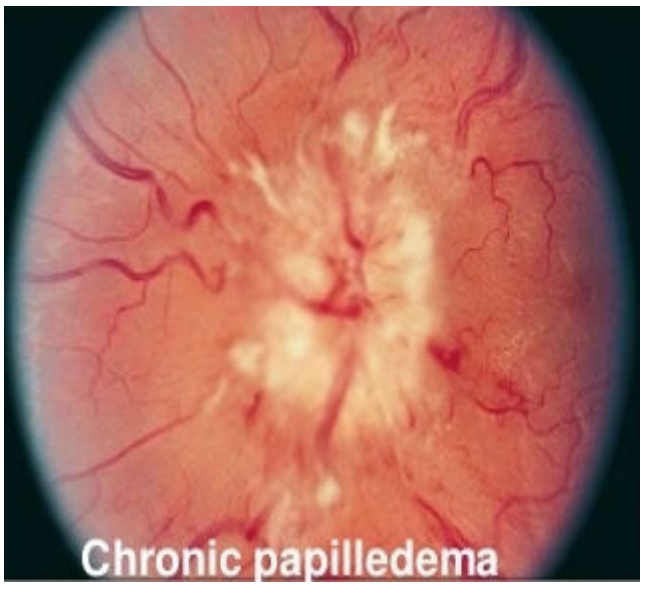
Established papilloedema

- Margins indistinct and cup obliterated
- Surface elevated upto more than +3 D with direct ophthalmoscope
- Flame-shaped haemorrhages, cotton-wool spots
- Venous engorgement and peripapillary oedema
- Paton's Lines-radial lines cascading from optic disc
- Macular star

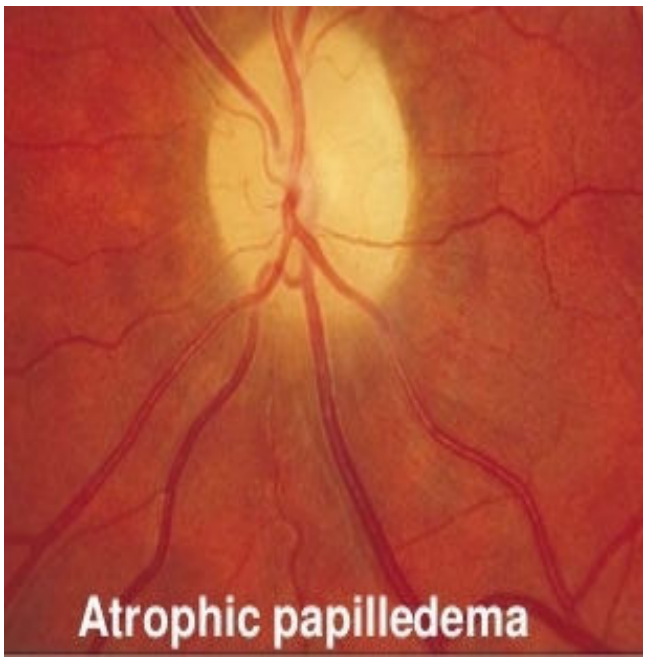


With progression

- Chronic papilloedema
  - Central cup remains obliterated
  - Haemorrhagic and exudative components resolve gradually
  - 'Champagne cork' appearance



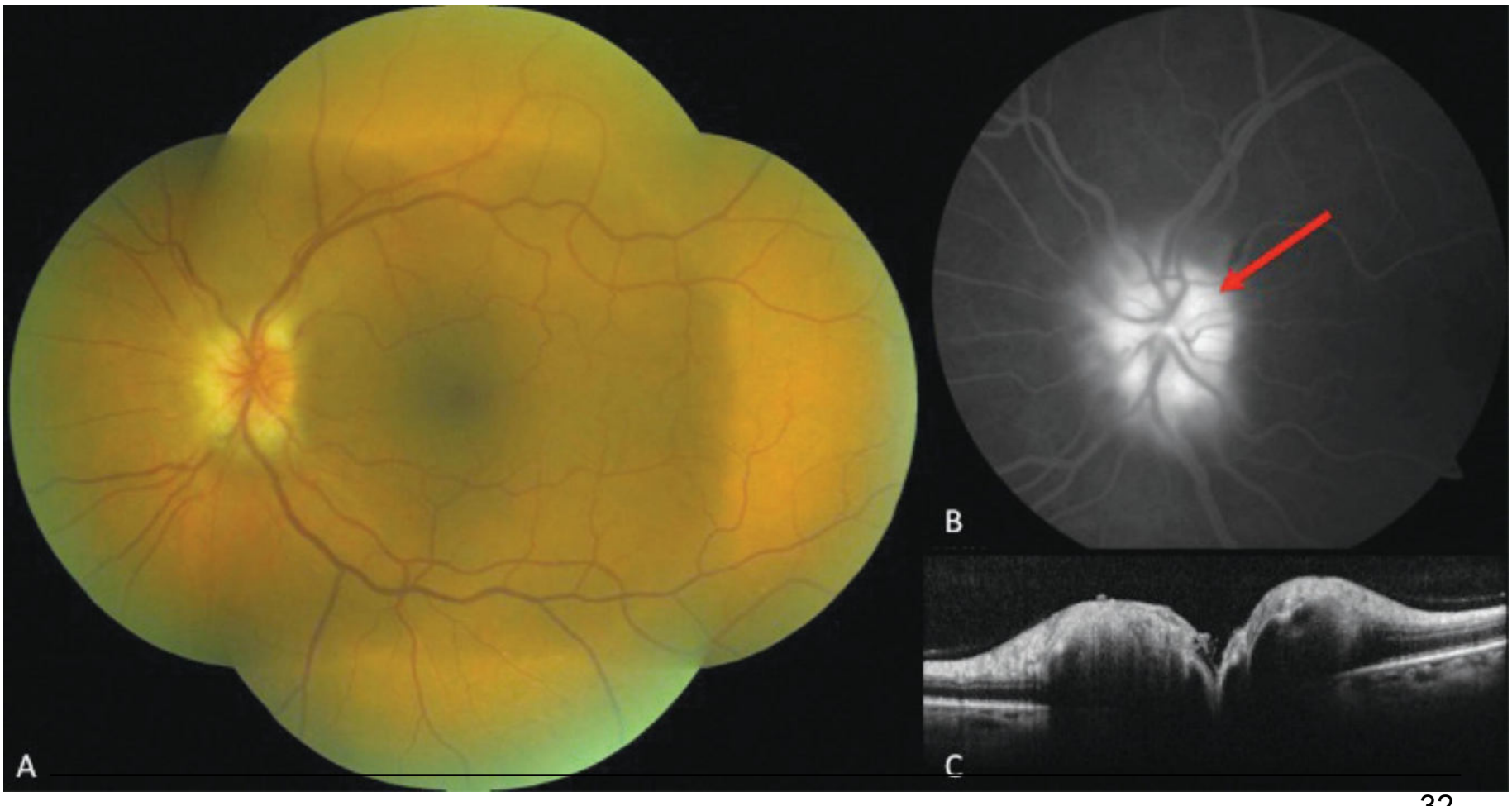
- Atrophic papilloedema
  - Retinal vessels attenuated with perivascular sheathing
  - Dirty white colour due to reactive gliosis
  - Leads to secondary optic atrophy



Visual fields

- Early-no changes
- Established stage- enlarged blind spot
- Chronic- peripheral constriction of field with nerve fibre bundle defects
- Finally- total loss of visual field

Fundus photo, FFA , OCT



	Papilloedema	Optic neuritis
History	Headache, vomiting	Rapid DV preceded by fever/respiratory infection
Laterality	usually bilateral	usually unilateral
VA	normal till late stage	severely reduced ≤6/60
Pain/tenderness of eyeball	absent	may be present
Pupil reaction	normal	RAPD (Marcus-Gunn's pupil)
Disc swelling	>+3 D in established	+2D to +3D
Haemorrhage, exudates	More, in established	relatively less
Visual fields	Enlarged blind spot, later gradual constriction	Central or centrocaecal scotoma
Colour vision	No effect	Affected
CT/MRI	SOL	Demyelinating disorder
Recovery of vision	May not be complete even after treatment	Usually complete after adequate treatment

Treatment

- Treat Cause
- Acetazolamide 250 mg 4 times a day
- Surgical decompression of optic nerve to preserve vision

Optic Atrophy

- Definition: Degeneration of optic nerve fibres with loss of their myelin sheaths characterised by pallor of the optic disc due to loss of vascularity owing to obliteration of disc capillaries

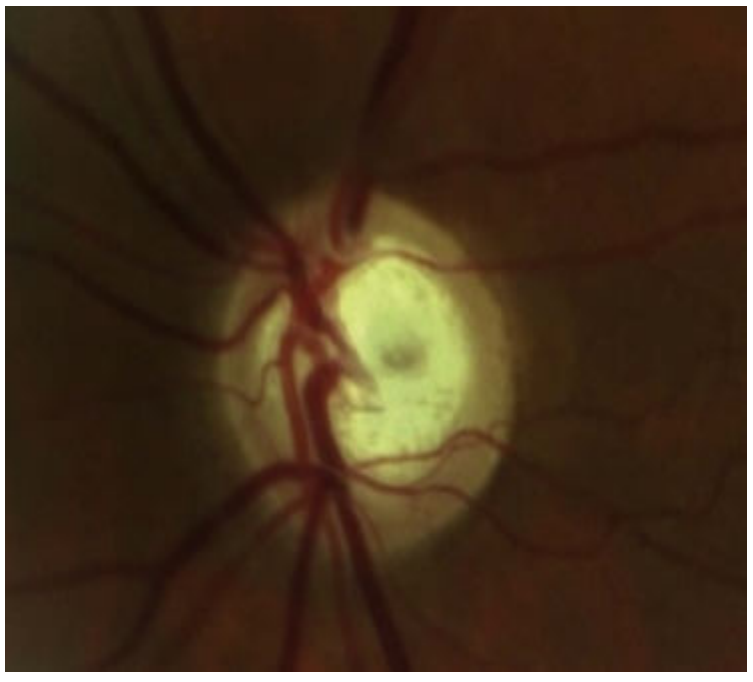
Classification-Aetiological

Primary Optic Atrophy

- No local disturbance, associated with CNS disease or no discoverable cause
- Commonest cause – Multiple Sclerosis
- Leber's optic atrophy
- Nerve compression: Tumour, hydrocephalus
- Injury to retrobulbar optic nerve

Cavernous type of Primary Optic Atrophy

- Deep excavated cup with undermined edges
  - Glaucomatous optic atrophy



Aetiological classification

- Secondary optic atrophy:

  preceded by swelling of optic disc- papilledema, optic neuritis, neuroretinitis
- Consecutive optic atrophy:

  follows extensive disease of the retina - Retinitis pigmentosa

  Long-standing retinal detachment

Anatomical classification

- **Ascending**: Lesion in retina, terminates at lateral geniculate body –
- Eg: RP, CRAO
- **Descending**: Disease involving optic nerve fibres anterior to LGB, terminates at optic disc –
- Eg: chiasmal compression

Symptoms

- Gradual/rapid loss of central/peripheral vision
- Impairment of colour vision

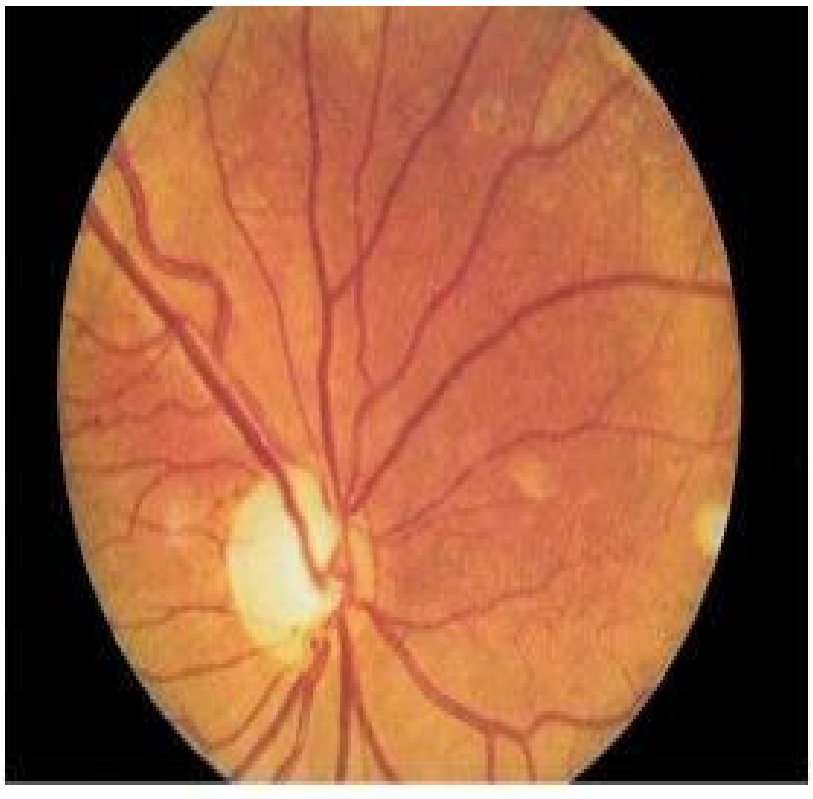
Signs

- Visual Acuity impaired in proportion to death of optic nerve fibres
- RAPD in unilateral Optic Atrophy

  Ultimately pupil dilated and immobile

Primary OA ophthalmoscopy

- Pale disc, classically paper white in colour
- Margins sharply defined
- Minimal atrophic cupping
- Blood vessels attenuated with marked reduction of small blood vessels on ONH to ≤6



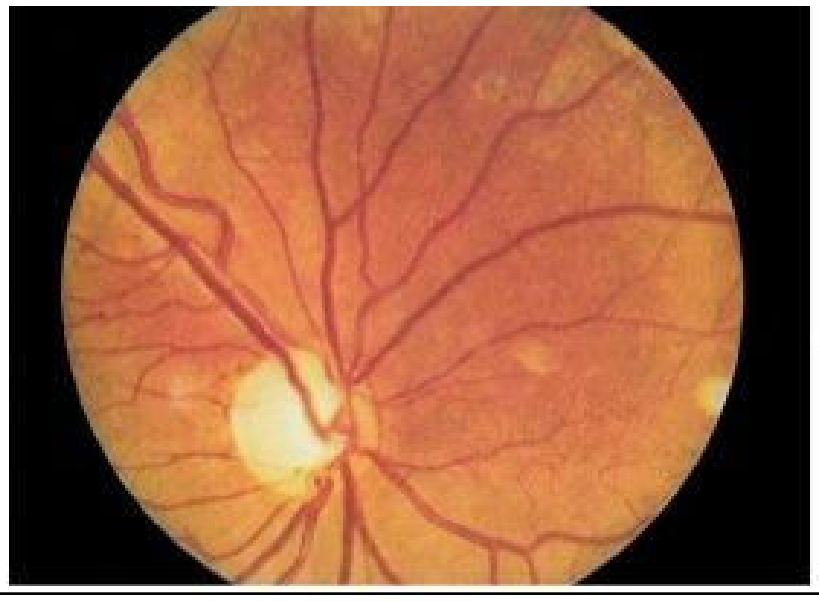
Primary Optic Atrophy

Secondary OA ophthalmoscopy

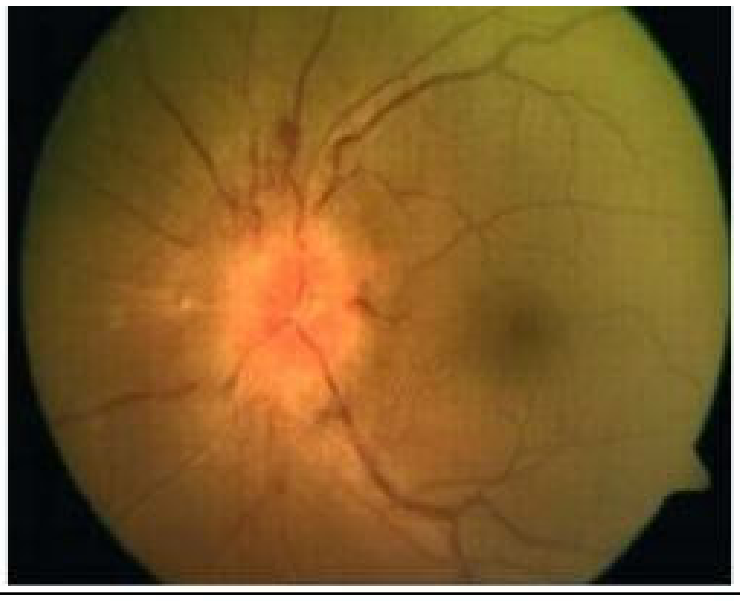
- Pale disc with dirty-grey colour, blurred margins
- Physiological cup is full, lamina cribrosa obscured
- Narrowing of blood vessels with sheathing
- Gliosis over disc surface extending towards peripapillary retina

Secondary Optic Atrophy

	Primary OA	Secondary OA
Appearance	chalky white	dirty grey
Margins	sharply defined	blurred
Cup	deep	obliterated
Laminar dots	visible	not visible
Glial proliferation	absent	marked
Vessels	no sheathing	sheathing
Previous disc oedema	absent	present



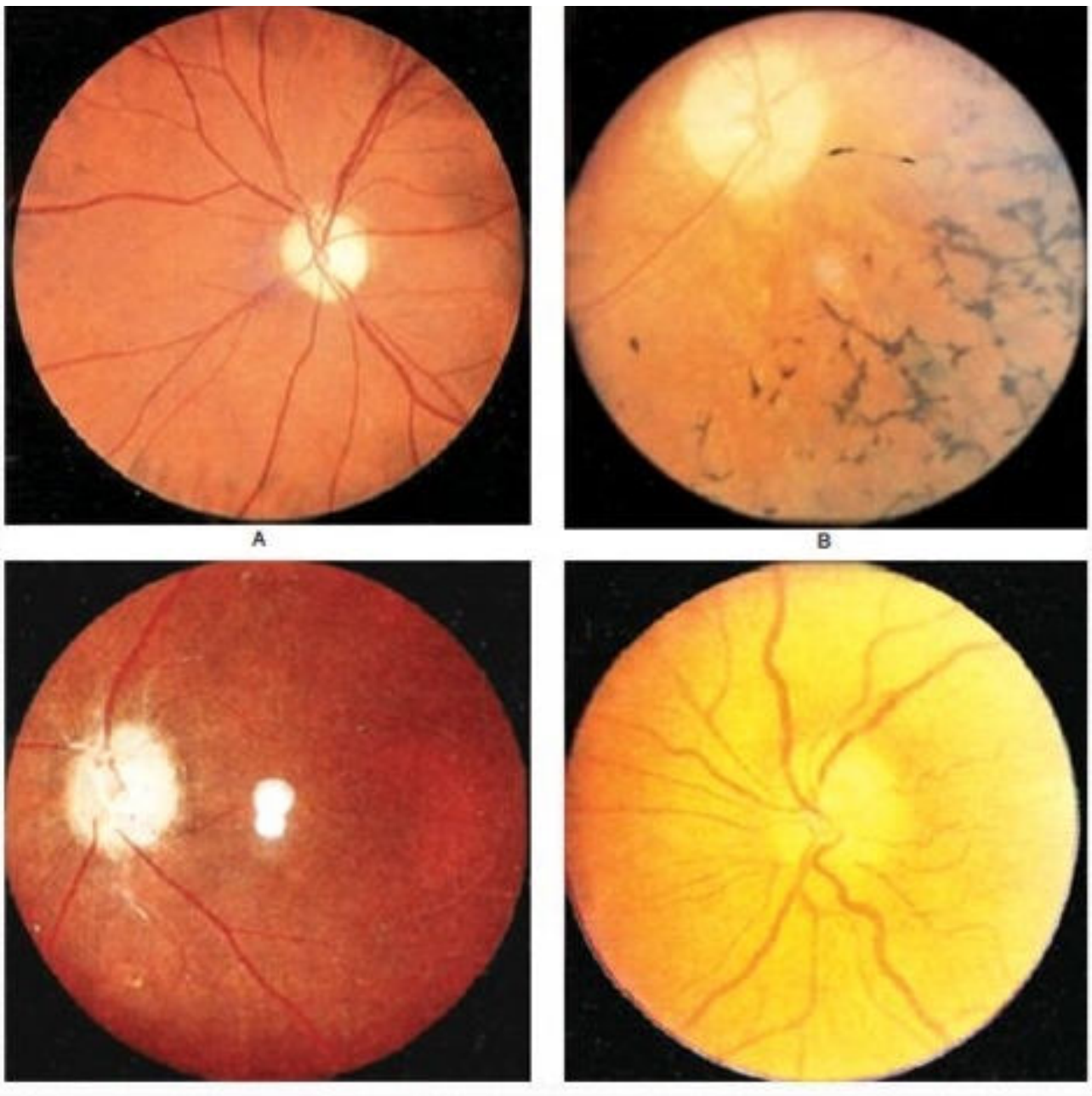
Primary Optic Atrophy



Secondary Optic Atrophy

Consecutive optic atrophy

- Yellowish-waxy pallor of the disc
- Margins less sharply defined
- Marked narrowing, even obliteration of retinal blood vessels



a) Primary, b) Consecutive, c) Postneuritic, d) Ischemic

Investigations

- Visual field: In partial OA, central vision is depressed with concentric contraction of the visual field
- FFA of Optic nerve head
- VEP especially in children
- Neurological evaluation

Hereditary optic atrophy

- Autosomal recessive/dominant
- LHON: Leber's hereditary OA

Treatment

- Treat the cause
- Gene therapy is emerging
- Community based rehabilitation in bilateral cases as prognosis is poor

Low-vision aids

Question

- The adjoining photograph belongs to a 12 year old boy who has difficulty in seeing at night.
- Can you identify the disc abnormality in the photograph?
- What condition does he suffer from?

Thank you