

بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

24 12:47AM

**RECOMMENDATIONS FOR
MANAGEMENT OF
OPTIC NEUROPATHY**

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NANOS**

International neuro-ophthalmology society INOS

Europien neuro-ophthalmology society EUNOS

OPTIC NEUROPATHY

- The causes of an optic neuropathy can be remembered by **NIGHT TICS**
- **N**euritis **I**schaemic **G**ranulomatous **H**ereditary

OPTIC NEURITIS

- Optic neuritis is inflammation of the optic nerve, caused by damage to and loss of the protective sheath (myelin) surrounding this nerve that is so vital for good vision. Demyelinating optic neuritis is another term for this eye condition
- . Less commonly, it can accompany other systemic inflammatory disorders such as systemic lupus erythematosus, syphilis, or sarcoidosis.

OPTIC NEURITIS

- Autoimmune disorders of the central nervous system often involve autoimmune inflammation of the anterior visual pathway
- Autoimmune optic neuropathy (AON), sometimes called autoimmune optic neuritis, may be a forme fruste of systemic lupus erythematosus (SLE) associated optic neuropathy.
-

OPTIC NEURITIS

- The spectrum of autoimmune optic neuropathies (ON) is extending. The phenotypic spectrum includes single isolated optic neuritis (SION)

Classification of optic neuritis

- **Ophthalmoscopic classification**
- Papillitis
- Retrobulbar neuritis
- Neuroretinitis
- **Aetiologic classification**
- Demylinating
- Parainfectuous
- infectius

OPTIC NEURITIS

*

Pain or discomfort around
the orbit **or with eye
movements**

*Decreased acuity is the
role

* Obliteration of
central cup

*Cells in the vitreous

*Deep retinal exudates
or macular star

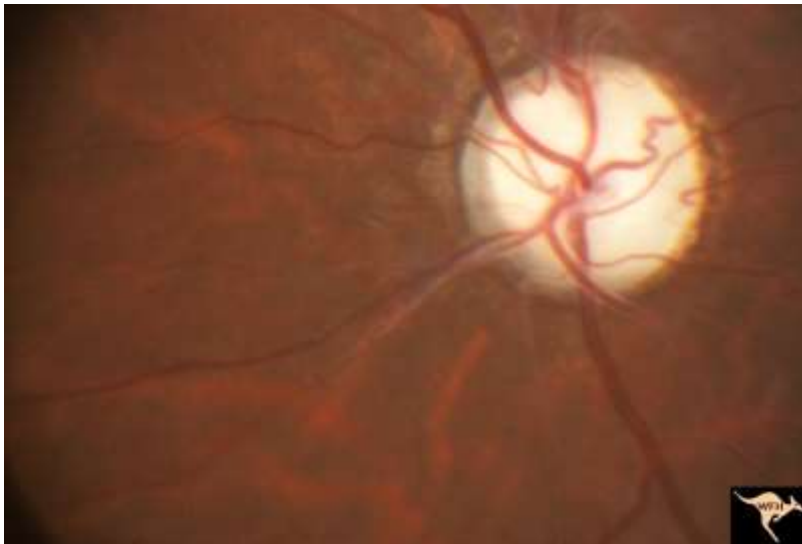


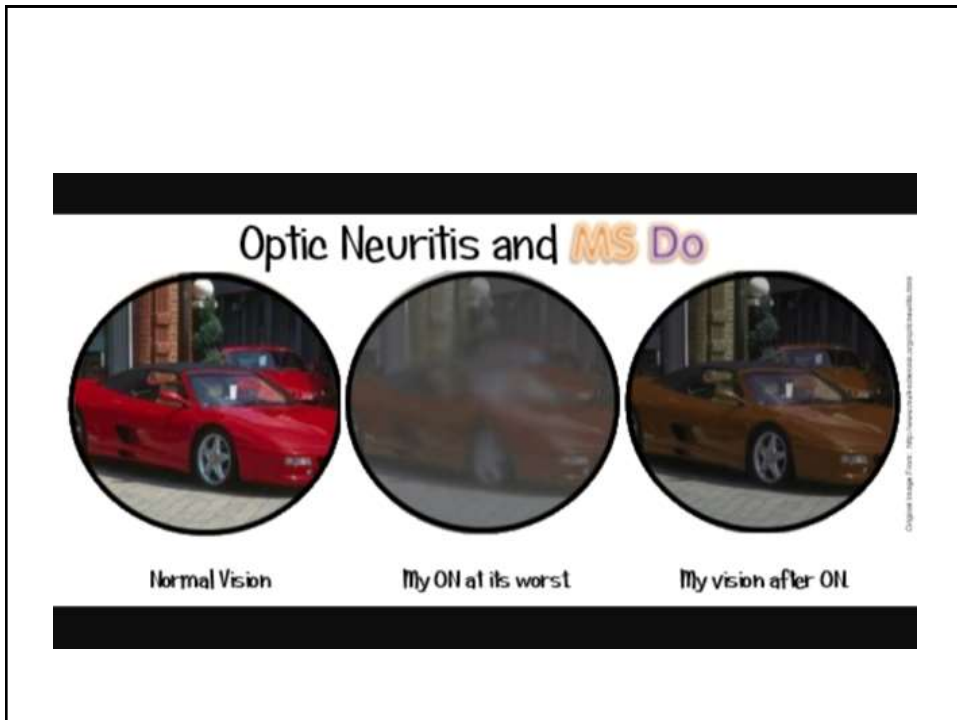
OPTIC NEURITIS

RAPD
COLOUR DEFICIT
UNPROPORTIONAL TO
THE DEGREE OF VISUAL
ACUITY LOSS
OPTIC DISC SWELLING IS
NOT CORRELATED TO
SEVERITY OF
DYSFUNCTION



POST PAPPILLITIC OPTIC ATROPHY





Retrobulbar neuritis

- **Retrobulbar pain on eye movements.**
- **Produce no ophthalmological visible changes in the disc.**

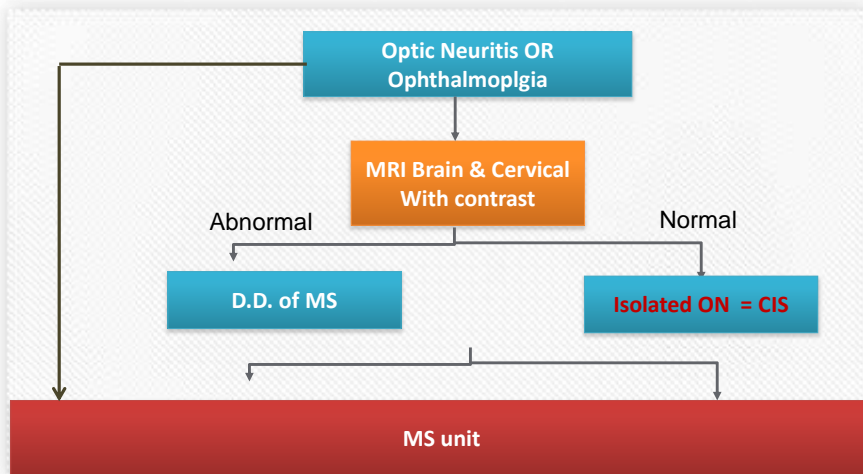
Chronic relapsing inflammatory optic neuropathy (CRION)

- Is a recently described recurrent optic neuropathy which is steroid responsive. Several features distinguish this entity from optic neuritis associated with demyelinating disorders and connective tissue diseases. The severe degree of visual loss, persistence of pain after onset of visual loss, and recurrent episodes are unique to this disorder.

ON ASSOCIATED WITH NMO(DEVIC,S)DISEASE

- NMO is an acute inflammatory demyelinating disease involving optic nerve and spinal cord
- NMO and MS identical in their initial presentation even NMO is more aggressive

Management of ON



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Optic Neuritis: lab workup

- Complete blood count (CBC)
- Serum vitamin B-12 and folate levels (eg, bilateral central scotoma)
- Lyme titers (eg, endemic area, tick exposure, rash of erythema chronica migrans)
- Tuberculin skin testing, chest radiography, or QuantiFERON-TB testing (eg, tuberculosis [TB] exposure, endemic area)

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- Fluorescent treponemal antibody (FTA) testing (eg, syphilis serology) or nontreponemal testing (eg, Venereal Disease Research Laboratories [VDRL] testing or rapid plasma reagin [RPR] testing)
- Antinuclear antibody (eg, systemic lupus erythematosus)
- HIV testing (eg, high-risk patients)
- Angiotensin-converting enzyme (ACE) level, chest radiography, lysozyme (eg, sarcoidosis)
- Erythrocyte sedimentation rate (eg, inflammatory disorders)
- Serum NMO antibody IgG (anti-aquaporin-4 [AQP4] antibody) testing

Optic neuritis treatment trial ONTT(recommendations)

Chest x ray, blood tests, and lumbar puncture are not indicated for typical cases of ON

Consider treatment of **MS** with intravenous steroids when 3-4 signals on MRI

Despite good visual outcome, there is damage of ON, nerve fiber layer thinning, and latency in VEP response

There is risk of recurrence in either eye in 10 years 35%, the risk is twice high in **MS** 48%

Good recovery despite axonal loss occurs due to redundancy in visual system or cortical plasticity

ONTT

- More than 90% recover in idiopathic ON
- **Immediate treatment:**
- 250 mg intravenous methylprednisone every 6 hours for three days followed by oral prednisone (1mg/kg/day) for 11 days with taper for 3 days ,then 15 days with 3 days taper .
- REFERRAL TO **MS** GROUP IS MANDATORY

PAPILLEDEMA

Passive edema of optic nerve head

Idiopathic increased intracranial pressure IIH

- Venous sinus thrombosis
- Space occupying lesion

Subdural hematoma, subarachnoid hge.

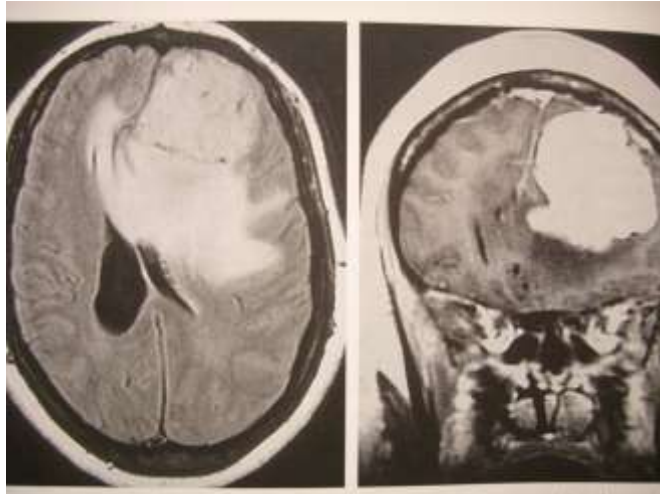
- Brain abscess, encephalitis, meningitis

Elevated intracranial pressure is transmitted to the optic nerve sheathes with resulting stagnation of venous return from retina and optic nerve head .

**Optic nerve fibers are compressed in the subarachnoid space resulting in disrupting intra axonal fluid mechanics with leak of water and protein into extracellular space of prelaminar portion of optic disc*

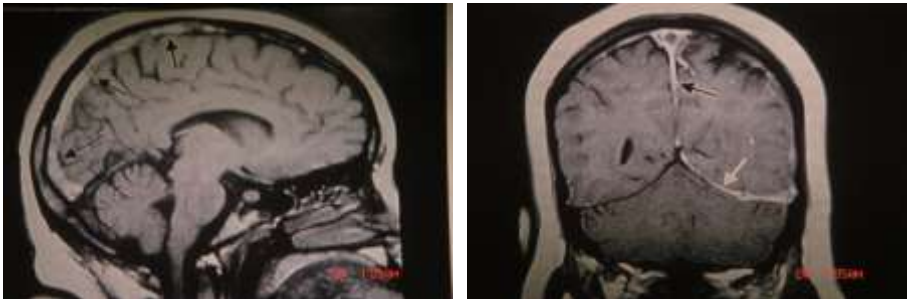
PAPILLEDEMA

*Space
occupying
lesion



PAPILLEDEMA

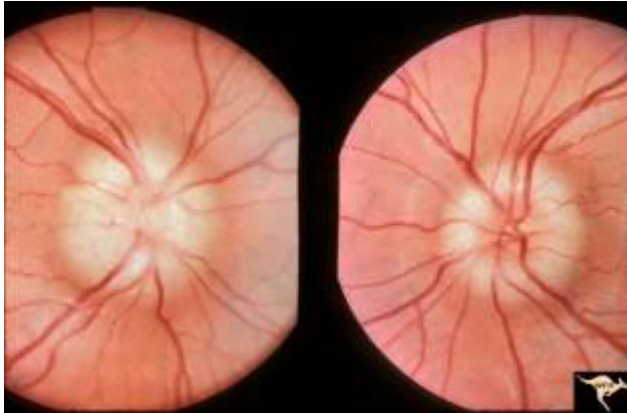
SAGITTAL & TRANSVERSE SINUSES THROMBOSIS
MRI,MRV



Treatment: Anticoagulants & Carbonic anhydrase
inhibitors & Antibiotics & Sinus stent

PAPILLEDEMA

IDIOPATHIC INTRACRANIAL HYPERTENSION I.I.H



Papilledema

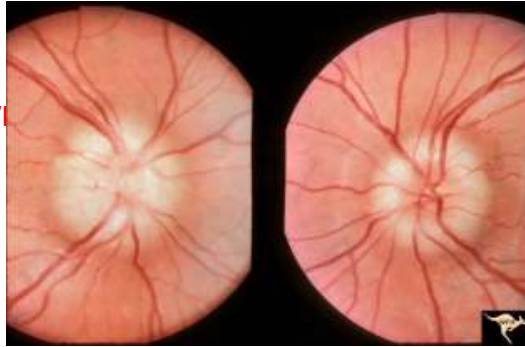
Idiopathic Intracranial Hypertension I.I.H

- **Look for Drugs: Antibiotics, tetracyclines, vitamin A, Contraceptive drugs.**
- **Pregnancy.**
- **Increased C.S.F. Opening pressure on lumbar puncture with normal composition**
- **CT scan is normal**
- **M.R.I. Very important to see distended sheath of optic nerves & exclusion of pituitary lesion and brain tumors.**

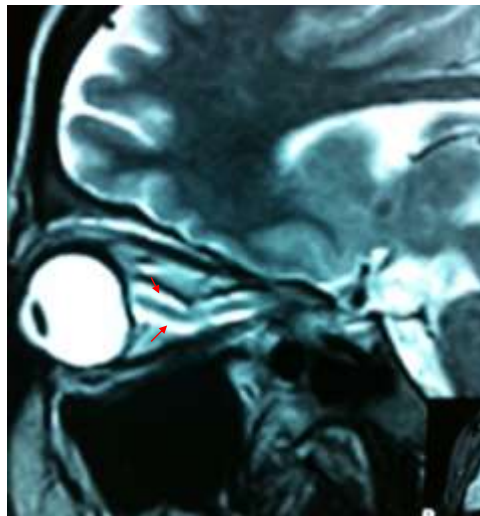
PAPILLEDEMA

IDIOPATHIC INTRACRANIAL HYPERTENSION **I.I.H**

- * HEADACHE, TRANSIENT VISUAL OBSCURATIONS
- * GRADUALLY DECREASED VISUAL ACUITY
- * DIPLOPIA 6th N. PALSY
- * BILATERAL DISC EDEMA & BLURRED MARGINS
- * VISUAL FIELD DEFECTS
- * NO VENOUS PULSATIONS



T2 MRI, distended optic nerve sheath



Papilledema

Idiopathic Intracranial Hypertension **I.I.H**

TREATMENT

Carbonic anhydrase inhibitors

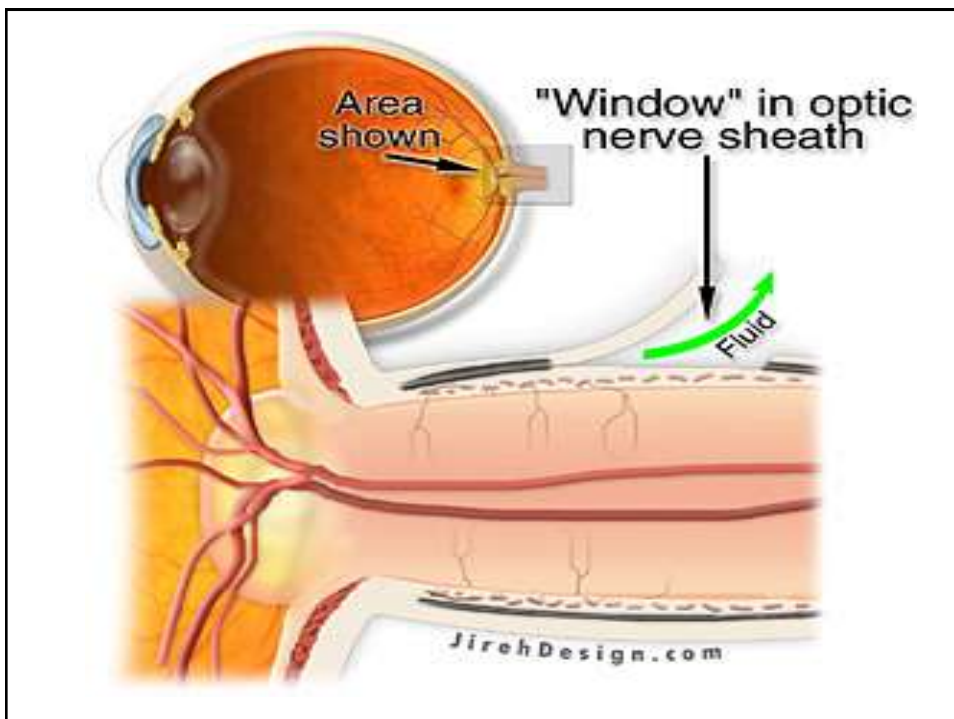
Diuretics

Lumbar puncture to release pressure and release papilledema.

If no improvement with deterioration of visual functions

optic nerve sheath decompression is indicated for one or both eyes according to severity and duration of papilledema.

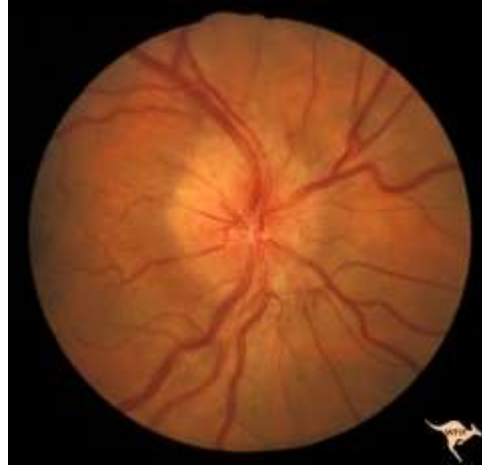
Lumboperitoneal shunt surgery is the procedure of choice if headache is severe



PSEUDOPAPILLEDEMA

CHARACTERISTICS

- Central cup absent but spontaneous venous pulsations
- Vessels arise from central apex of disc
- Increased number of major disc vessels
- Disc margins irregular with deranged peripapillary retinal pigment epithelium
- No haemorrhages
- No exudates or cotton wool spots



Papilledema or Pseudopapilledema?

Elevated optic nerves + headaches =
Increased ICP

Ultrasound, 30 degree test

CT

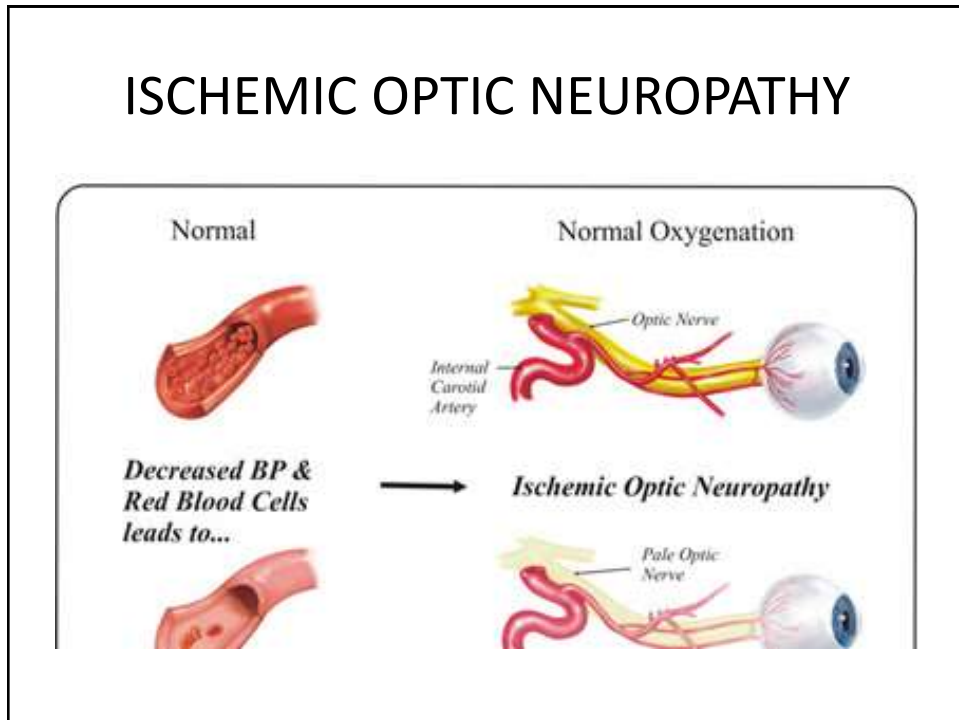
Red-free photos (surface drusen)

Stereo disc photos

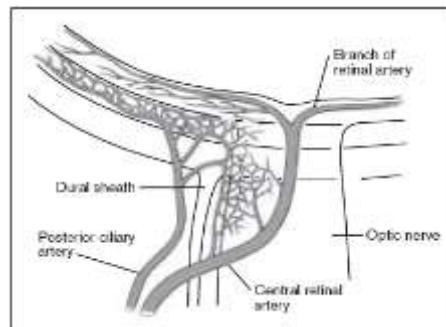
OCT



ISCHEMIC OPTIC NEUROPATHY



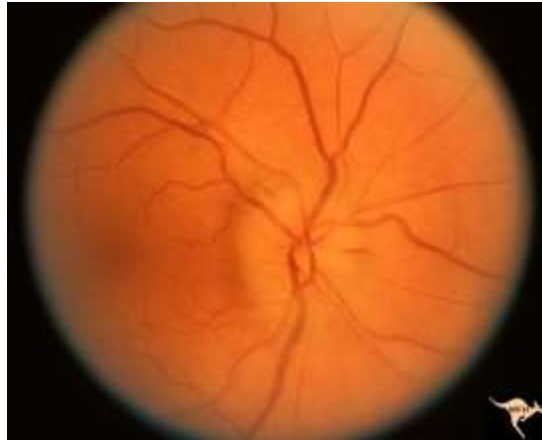
ANTERIOR ISCHEMIC OPTIC NEUROPATHY AION



- Blood supply of optic nerve head:
- *Retinal nerve fiber layer by CRA or cilioretinal.
- *Prelaminar region by centrepital peripapillary choroidal v.
- *Laminar region by centrepital sh.post. Ciliary arteries.
- *Retrolaminar region by recurrent pial branches from peripapillary choroidal vessels .

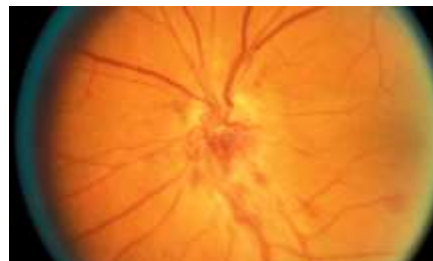
- Blood flow to optic N. head depends on:
 *B.P. & *I.O.P
 &*Vascular resistance.
- Resistance to blood flow depends on Autoregulation, if damaged the optic nerve becomes susceptible to ischemia
- Subclinical ischemia with failure of autoregulation cause axoplasmic stasis leading to disc swelling

NON ARTERITIC AION



NAION

- This axoplasmic swelling within the restricted space of **disc at risk** compress the nutrient capillaries with further ischemia ending with clinical disc hyperemia and nerve fiber layer haemorrhages.
- **Systemic risk factors for NAION**
 - Hypertension ,diabetes,
 - ischemic heart disease,
 - cerebrovascular accidents,atherosclerosis,peptic ulcers.



NAION

- **Ocular risk factors for NAION:**

small optic nerve head, increased number of branches of CRV on the disc,*abundant appearing nerve fiber bundle layer with heaping along the superior ,inferior, nasal borders

DISC AT RISK .

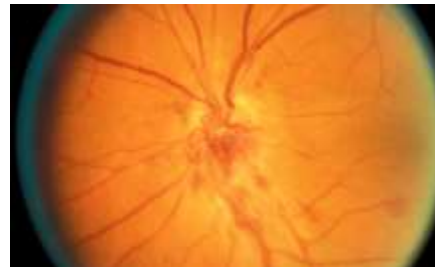
Hypertropia

Elevated I.O.P.



NON ARTERITIC AION

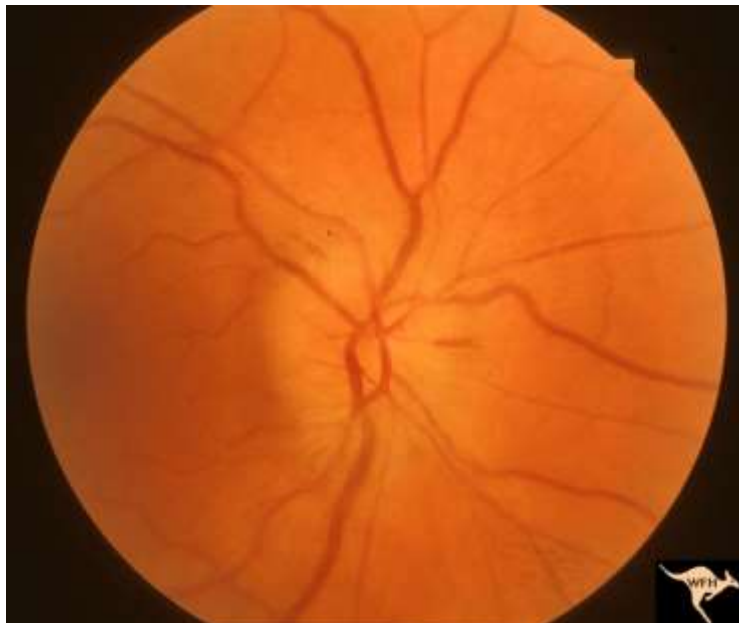
- Both sexes equally affected
- Age range 40-80 mean 55
- Bilateral in 30% in 3-5 years in young & diabetics
- Sudden painless loss of V.A. in one eye discovered on awakening up in the morning.
- Progressive loss of visual field over days or weeks . Inferior altitudinal field defect very common 70- 80%
- Afferent pupillary defect and diminished colour perception



NAION

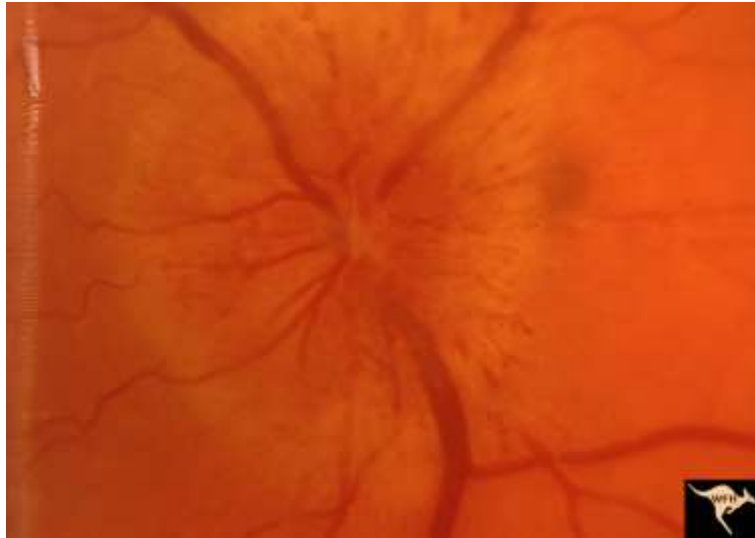


NAION



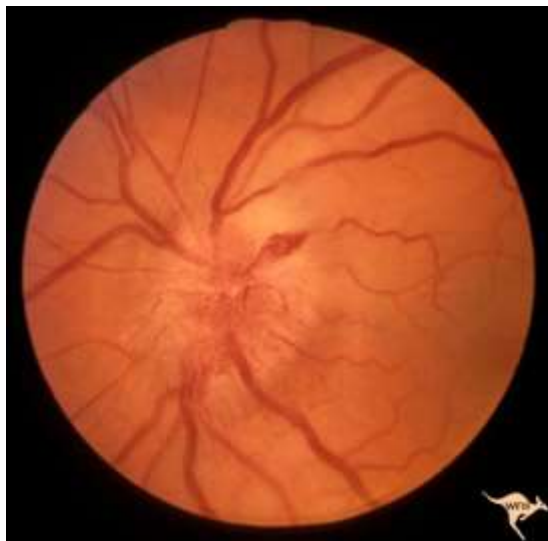
NAION

*Edema is mild to moderate with leakage on F.A.
 *Sectorial pallor either superior or inferior corresponding to altitudinal field defect



NON ARTERITIC AION

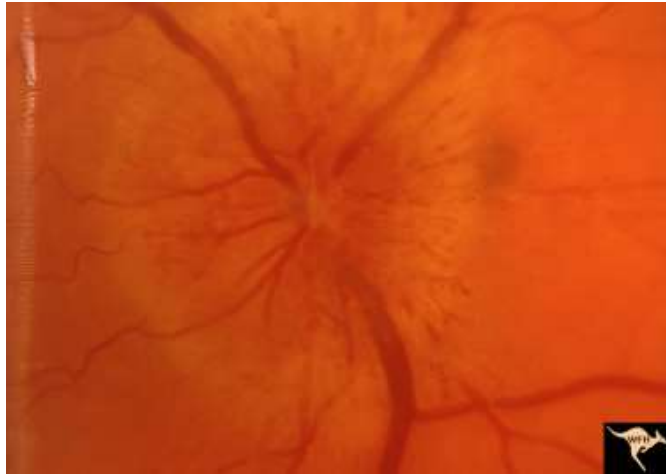
- ***Pallid edema** of optic disc with papillary or peripapillary nerve fiber layer haemorrhages and exudates distinguish it from papillitis in middle aged people



NAION

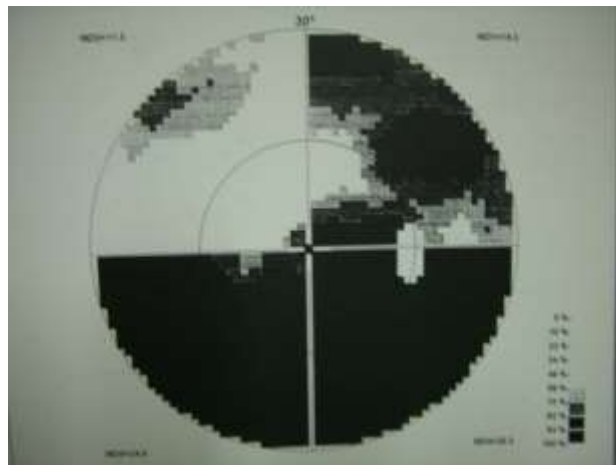
Edema is mild to moderate with leakage on F.A.

**Sectorial pallor either superior or inferior corresponding to altitudinal field defect*



NAION

- Inferior altitudinal visual field defect is very characteristic in 70%
- Of cases



Treatment of NAION: there is no proven efficient treatment for NAION

Several treatment have been tried:

- *Corticosteroids
 - * Hyperbaric oxygen therapy
 - *Levodopa and carbidopa
 - *Osmotic diuretics
 - *Treatment of hypertension &diabetes
 - *Vasodilators & neurotonics
 - *Role of anti VGEF (AVASTIN)**FAILURE**
- ERYTHROPOITEN INTAVITREAL INJECTION**

AION

Arteritic AAION Giant cell arteritis

- **Systemic necrotising vasculitis of medium and large arteries.**

Age group 75 years



ARTERITIC AION

Infarction within
prelaminar and
laminar optic nerve
due to vaso-
obliterative
occlusion of short
posterior ciliary
vessels



ARTERITIC AION

Diplopia, headache
Scalp tenderness,
jaw claudication
Abnormal
superficial
temporal arteries
painful indurated
prominent and
without pulse



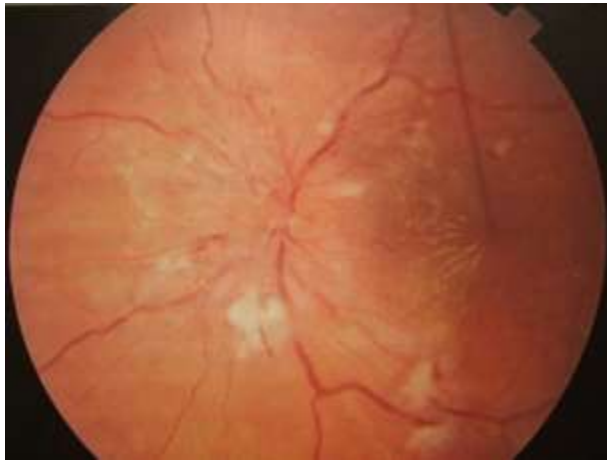
AAION

- Mild disc swelling with advanced pallor
- Delayed filling of dye on F.A.
- Beading of vessels due to involvement of retinal artery circulation and post. ciliary arteries
- Prove diagnosis by temporal arter biopsy.
- **Treatment** by high doses of intravenous or oral corticosteroids often for prolonged time to preserve vision.



Hypertensive optic neuropathy

- Bilateral optic disc swelling in hypertensive patients
- Decreased visual acuity
- constricted field
- RAPD
- hypertensive fundus

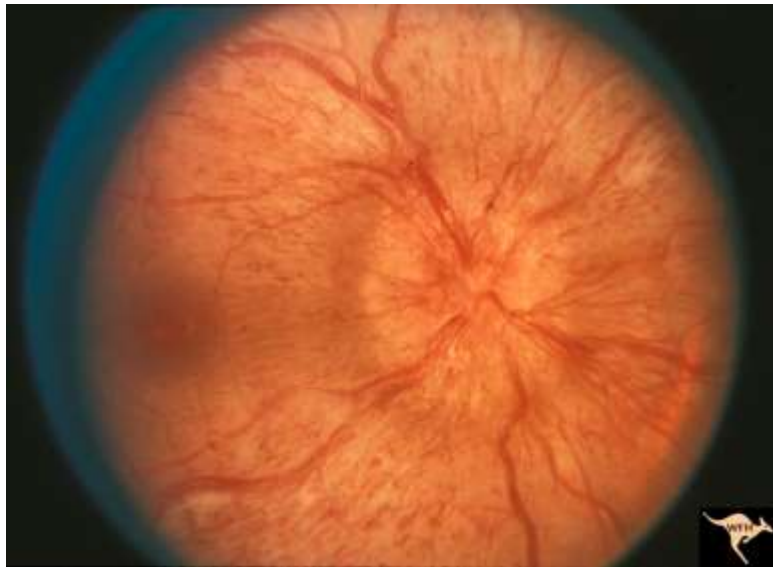


Diabetic papillopathy

- Atypical form of NAION
- Visual loss
- Optic disc swelling with peripapillary haemorrhages more than in NAION
- RAPD
- Diabetic retinopathy



Diabetic papillopathy



Toxic optic neuropathy Tobacco& Alcohol

- Nausia , vomiting, respiratory distress, headache, visual loss.
- Pallor and cupping of the disc
- Pupil sluggish then dilated fixed
- Very bad prognosis
- New treatment trials with success rate with:
- **ERYTHROPOITEN** injection intravenous and intravitreal

Sarcoid optic neuropathy

- Multisystem granulomatous ocular, neurologic, ophthalmic manifestations
- Slowly progressive decreased vision
- RAPD
- Anterior granulomatous uveitis, retinal ,choroidal lesions, lids, lacrimal glangs
- Pulmonary functions
- Corticosteroids

OPTIC NERVE GLIOMA

- Benign tumor of optic nerve with neurofibromatosis
- RAPD, Proptosis, ocular motility disturbances,
- Optic disc swelling.
- Fusiform mass in MRI
- Chemotherapy before 5 years
- Radiation therapy after 5 years

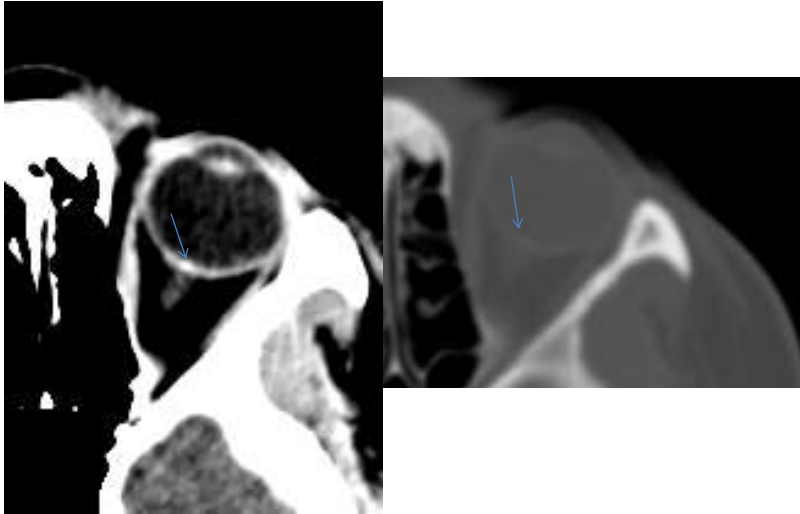


Optic disc drusen

- Accumulation of hyaline material within optic nerve that appears glistening.
- CT scan , Ultrasonography
- Can cause visual field defects
- No therapy is effective



CT showing optic disc drusen (still seen in bone window “calcified”)



OPTIC DISC GRANULOMA
(CAT SCRATCH DISEASE)

