

optic nerve functions

1. Visual Acuity
2. Color Vision
3. Pupil
4. Contrast sensitivity

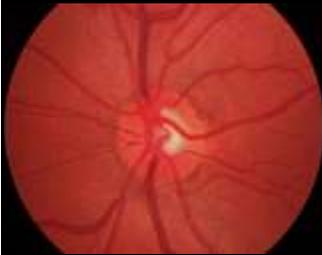
Ancillary Tests

1. Visual Field
2. Neuro-imaging
3. OCT
4. VEP

Optic nerve diseases

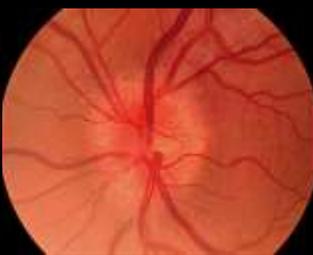
1. inflammation: optic neuritis
- 2 . ischemic optic neuropathy
- 3-Compression
- 4-Granuloma & infiltration
- 5-Hereditary
- 6-Toxic
- 7-Irradiation
- 8- Trauma

Optic Neuritis



1-Retrobulbar neuritis

optic disc is normal
in adults
with multiple sclerosis.



2-Papillitis

hyperemia and edema
common in children.



3-Neuroretinitis

Papillitis
with macular star
least common type
viral infections

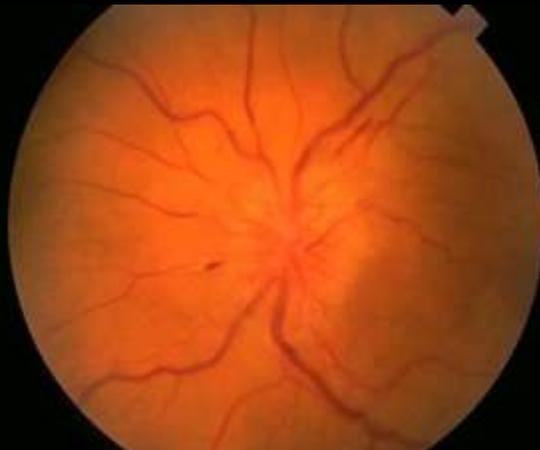
Rapid unilateral loss of vision

RAPD

Loss of color vision

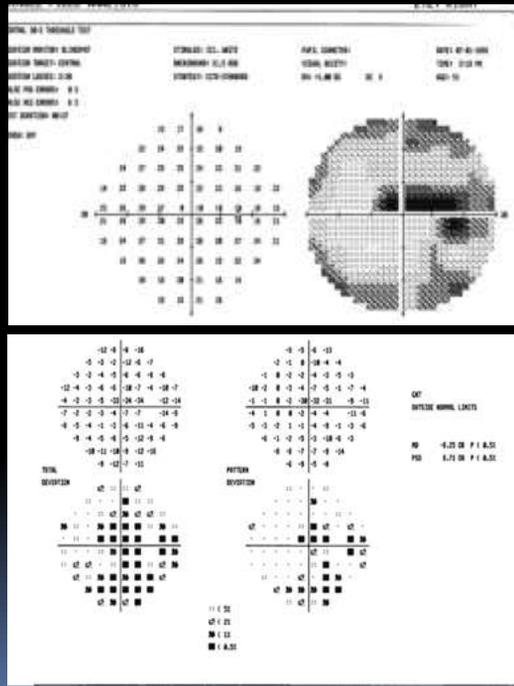
Pain in moving
the eye

**Swollen disc
with or without
peripapillary
flame-shaped
hemorrhages.**



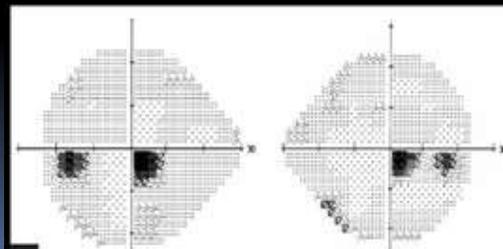
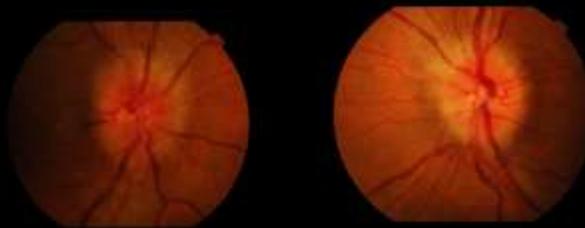
optic neuritis

Centrocecal scotoma

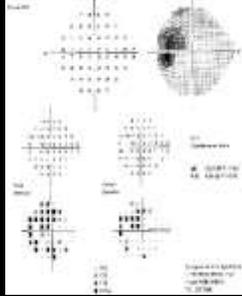


Bilateral optic neuritis

Bilateral Central scotoma

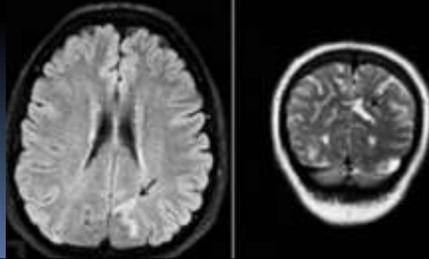


Bilateral hemianopsia



MRI

demyelinating lesions
multiple sclerosis

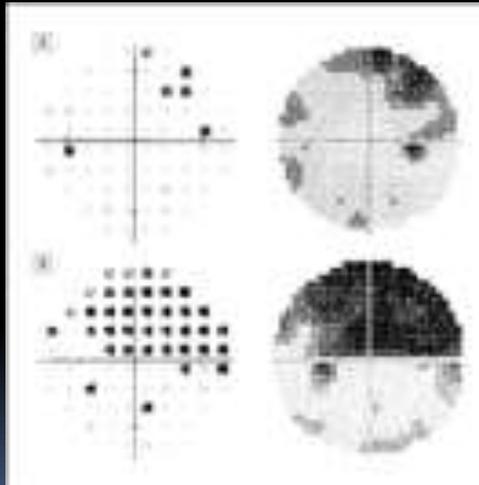


Neuromyelitis Optica

VF showed non-central scotoma

altitudinal VF

an ischemic mechanism play a role in ON in NMO patients

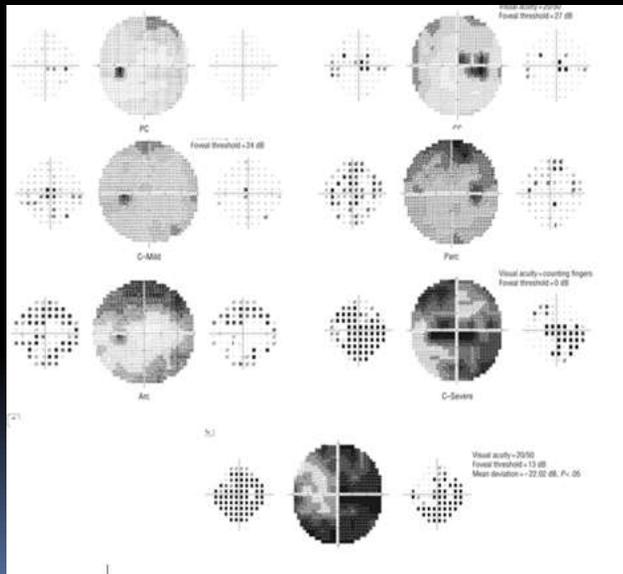


Optic Neuritis Follow Up

Diffuse and central loss in the affected eye at baseline

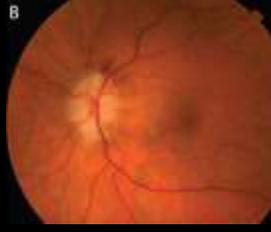
Follow Up :
nerve fiber bundle defects were the predominant localized abnormalities in both the affected and fellow eyes

physicians evaluate the characteristics of optic neuritis and other optic neuropathies in the future



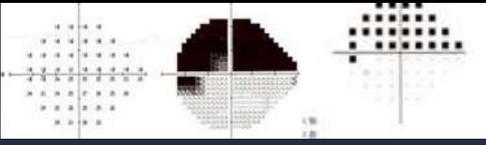
Ischemic optic neuropathy (ION)

NAION



B

sudden, painless visual loss
OS shows AION .

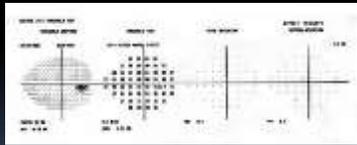


Upper altitudinal loss OS.



A

OD crowded disc



Normal VF OD

painless loss of vision

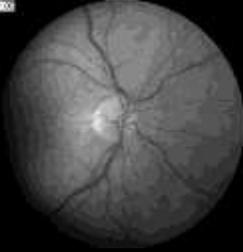


disk edema
peripapillary hemorrhages.

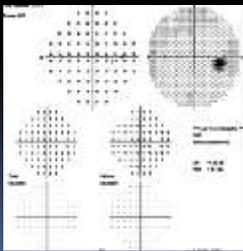


inferior altitudinal defect

OD shows initial visual acuity of 20/20



small optic disk
cup to disk ratio < 0.1
classic "disk at risk",

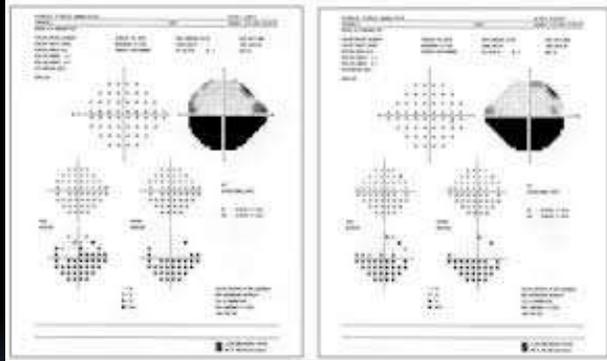


Normal VF

BILATERAL NA-AION FOLLOW UP

Bilateral inferior altitudinal defect

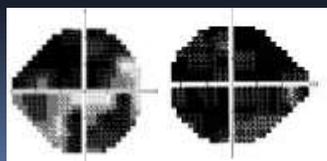
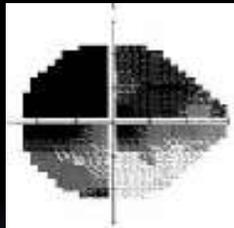
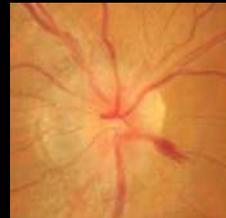
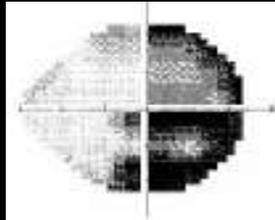
1. VF at baseline
2. every 2 weeks till resolution of disc edema
3. then monthly till the fields stabilize (2- 6 month).



The normal fellow eye has a 15% risk of NAION within five years

NAAION

- OD day 1
- OS day 11
- OU day 90

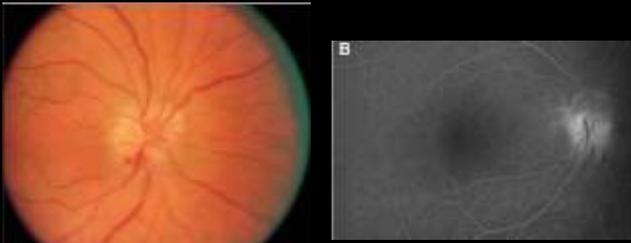
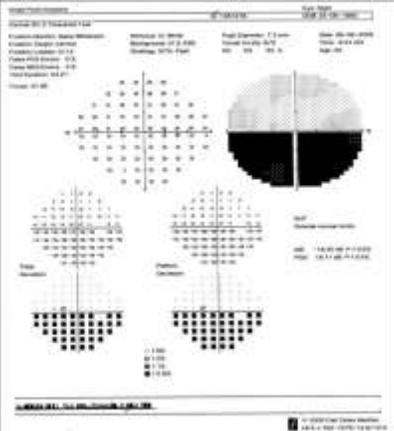


1 st. attack

At presentation
inferior ONH edema

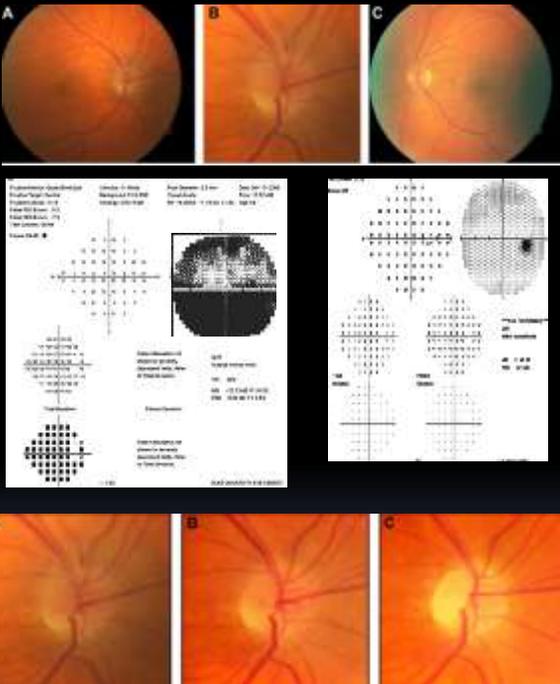
FA :
leakage of the
inferior ONH.

VF :
Inferior altitudinal
field defect

- OD: after 2 weeks shows superior ONH edema.
- OS: ONH is normal, crowded.
- VF of OD :
Upper arcuate defect and stable inferior altitudinal defect.

2 month pale ONH



Non-arteritic AION

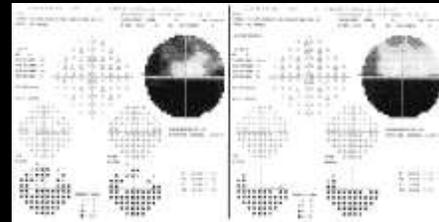


diffuse or sectorial edema•
Few splinter hemorrhages•

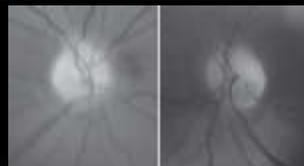
Resolution of edema and hemorrhages•
Optic atrophy and variable visual loss•

Bilateral posterior ischemic optic neuropathy (PION) after lumbar spine surgery

VF : 2 weeks postoperatively

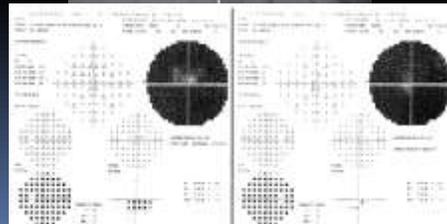


Optic disc : 2 months postoperatively
bilateral pallor



VF : 2 months postoperatively

C/O visual loss in the postoperative period
normal fundus and normal Neuroimaging
diagnosis of PION
close follow-up is mandatory.



PAPILLEDEMA

Etiology

1. Pseudo-tumor cerebri.
2. Space-occupying intracranial lesions.
3. Microbial infection: meningitis, encephalitis.
4. Hypertensive encephalopathy
5. Intracranial vascular lesions: sub-arachnoid hemorrhage, sagittal sinus thrombosis, etc.
6. Head-injury - long standing contusions, carotid-cavernous fistula
7. Metabolic disorders: Addison's disease, hypo-parathyroidism, etc.
8. Blood dyscrasias : leukemia, polycythemia, etc.
9. Choroidal plexus tumour by hypersecretion.
10. Miscellaneous; serum sickness, status epilepticus, sarcoidosis

PAPILLEDEMA



bilateral and asymmetric .
Hyperemic ONH.
Loss of ONH margins

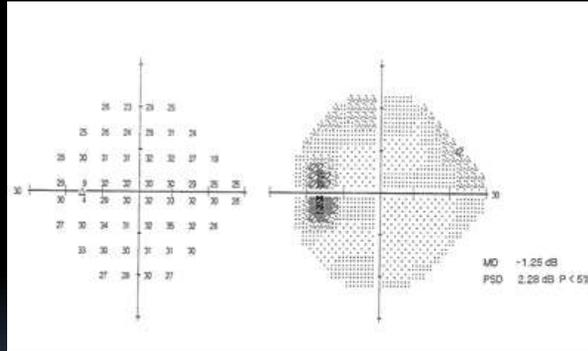


Flame-shaped hemorrhages
Disturbed axoplasmic flow

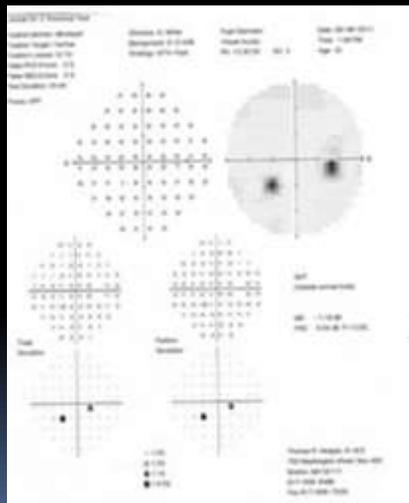
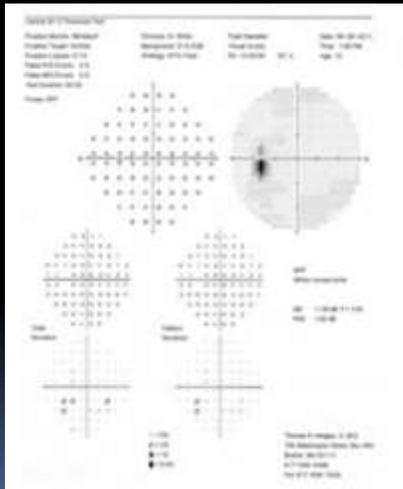
Papilledema

Enlarged blind spot

is a visual field defect in papilledema



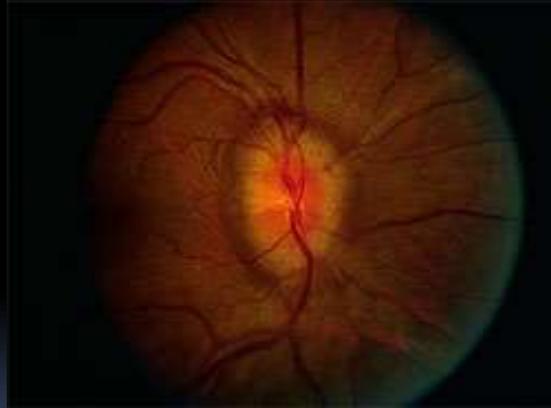
VF 30-2 mild bilateral enlarged blind spot



Pseudotumor Cerebri , idiopathic intracranial hypertension (IIH)

patients with signs and symptoms of increased intracranial pressure (ICP) of unknown cause

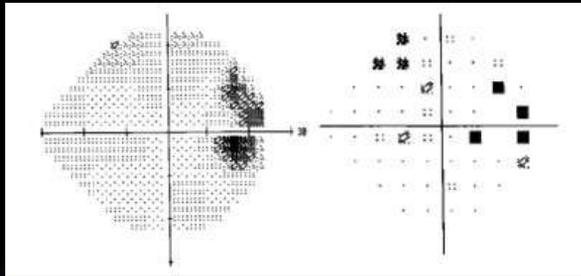
poor absorption of CSF by meninges

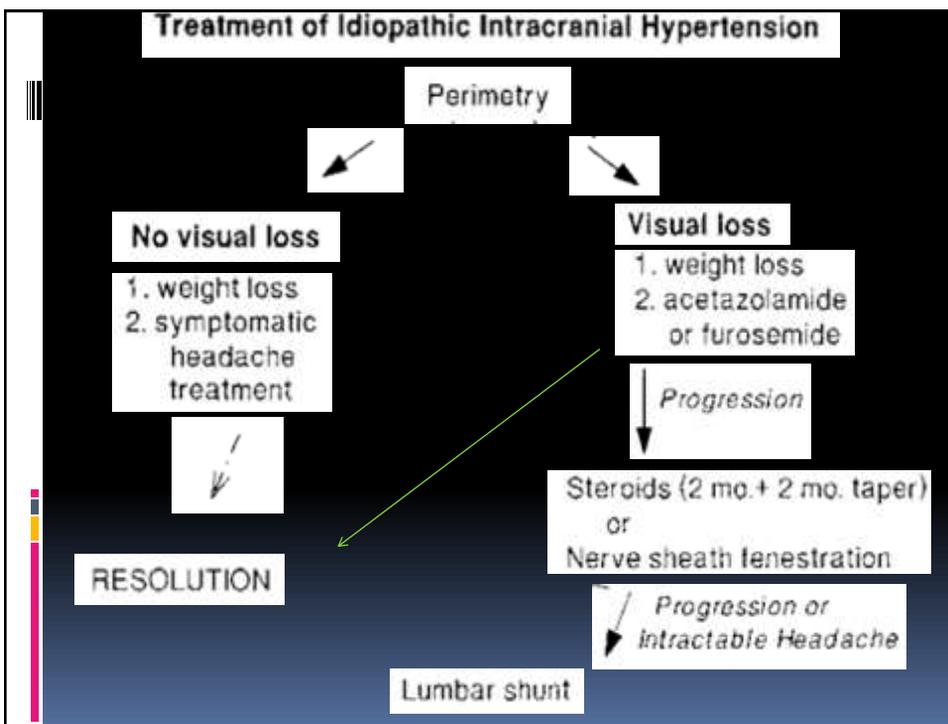
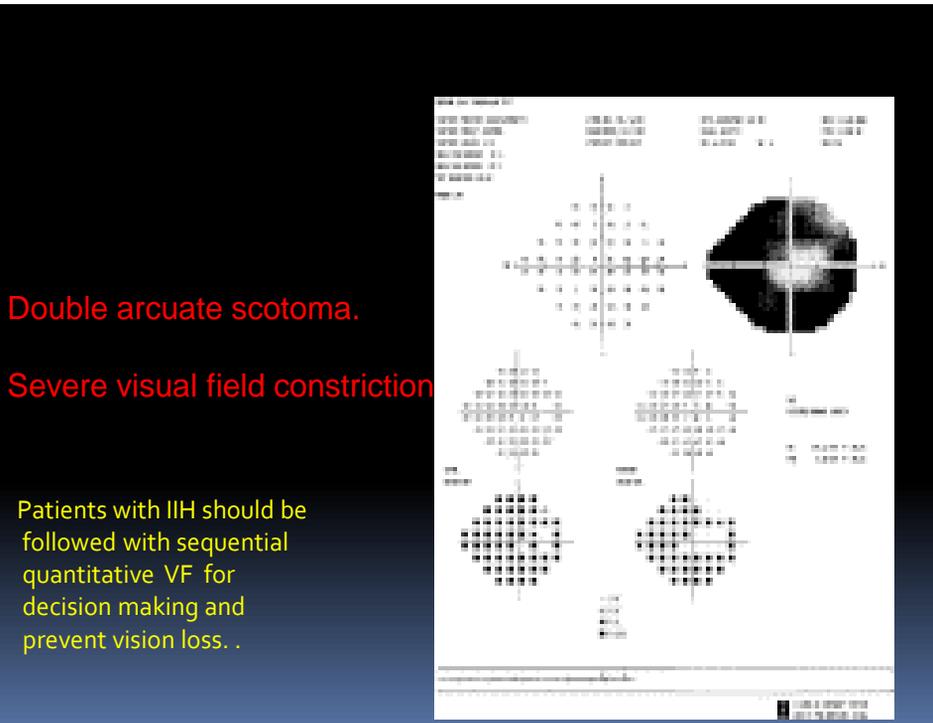


IDIOPATHIC ICH

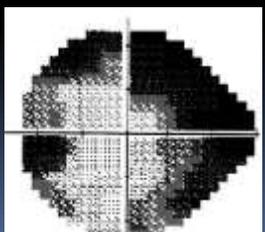
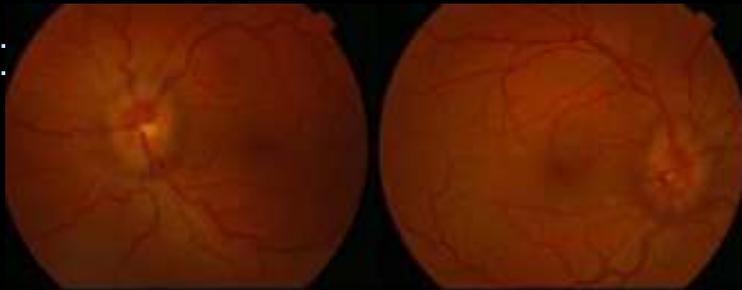
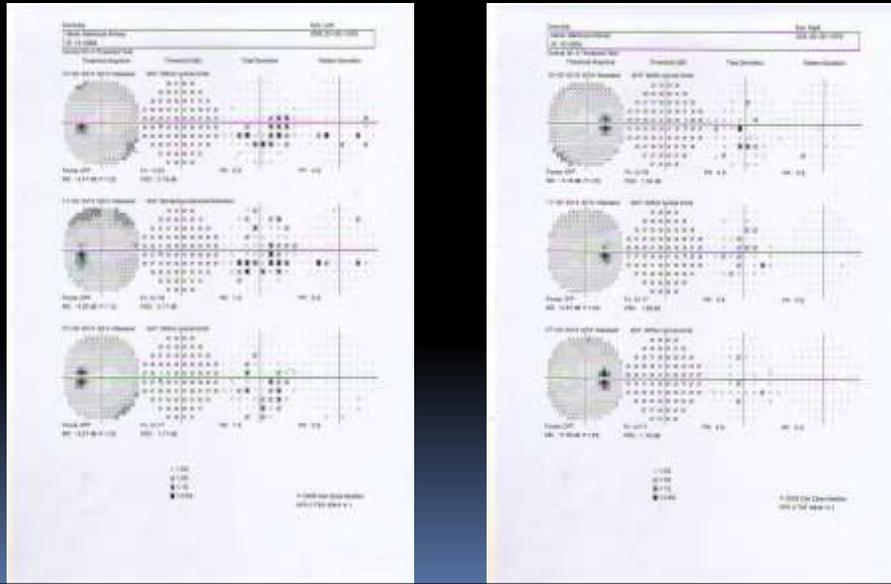
VF :
in idiopathic intracranial hypertension.

Enlarged blind spot

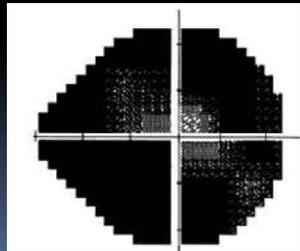




VF follow up in Papilledema Under medications



Decision for surgery

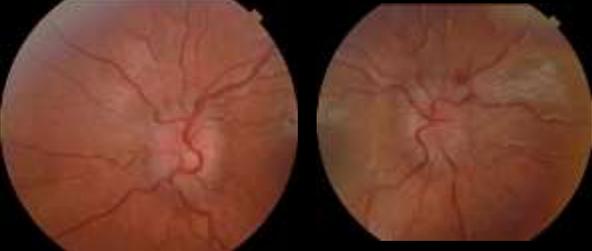


Medulloblastoma

Children brain tumor

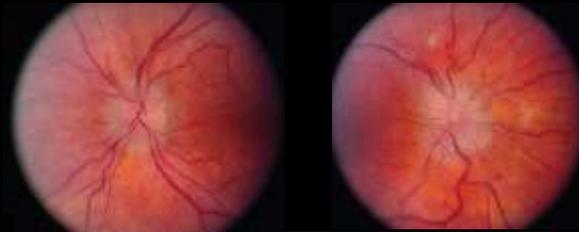
Bilateral papilledema due to increased ICT

disc edema
peripapillary splinter hemorrhage
superonasally

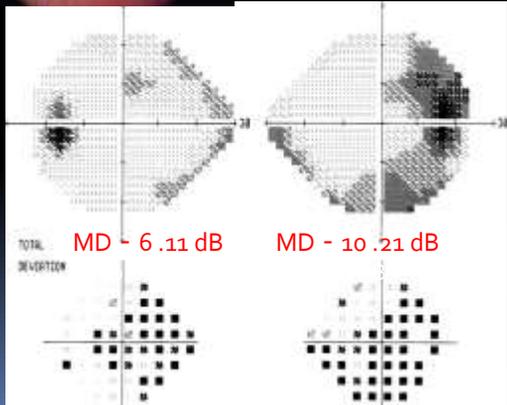


brain stem medulloblastoma

Fundus :at presentation
optic disc edema
bilateral (papilledema)



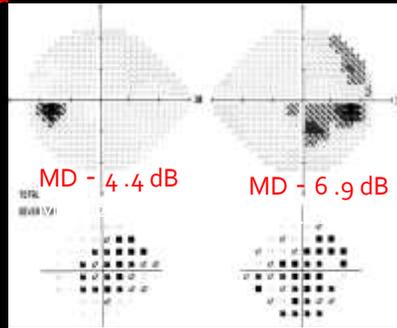
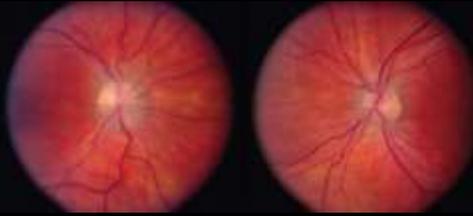
VF :
bilateral nerve fiber
bundle loss .



TOTAL DEVIATION MD - 6.11 dB MD - 10.21 dB

Brain stem medulloblastoma

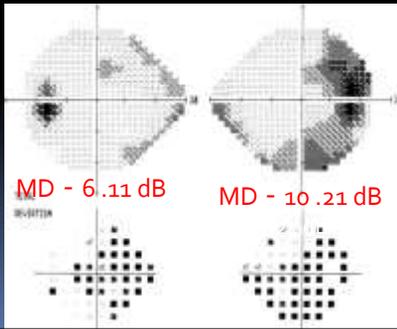
3 months after resection
of the tumor
resolution of papilledema.



VF : 3 months after surgical resection

shows improved VF

MD
-6.9 dB OD
-4.4 dB OS

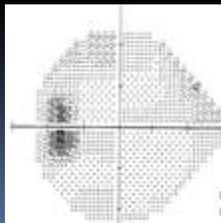


papilledema

visual acuity ,RADD
are normal.

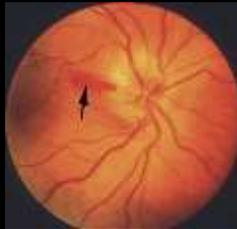


Bilateral disc edema
Splinter hge ,
Obliterated cup

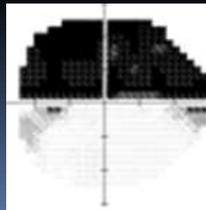


NAION

Sudden painless loss
of vision



optic disc edema
hyperemia,
splinter hge.

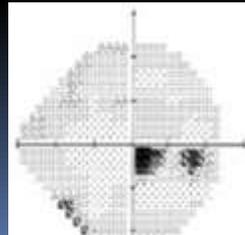


Optic neuritis

Painful Loss of vision
RAPD
Color vision defect



optic disc edema
peripapillary hge



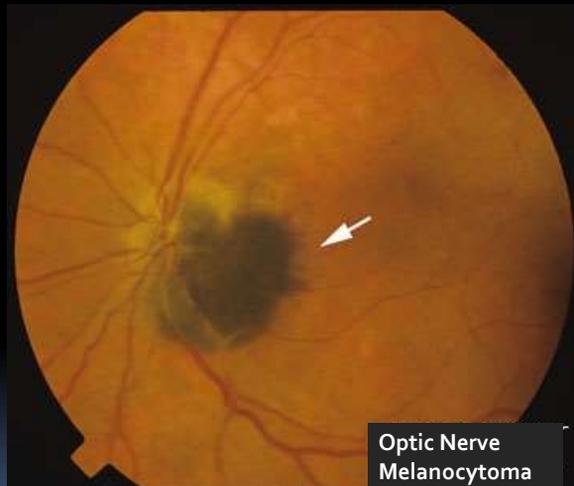
Optic nerve head tumors

Optic Nerve Melanocytoma

benign tumor made up of melanocytes and melanin

slowly grow

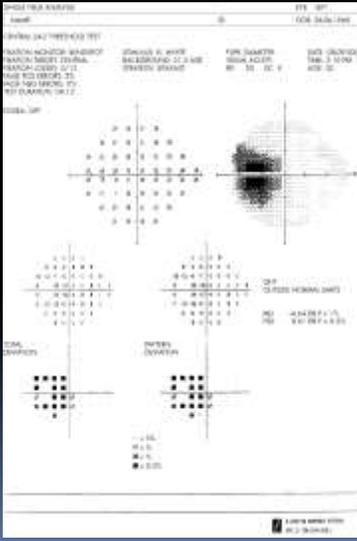
extend onto the surface of the optic nerve and invade the nerve fiber layer (feathered edge).



Optic Nerve Melanocytoma

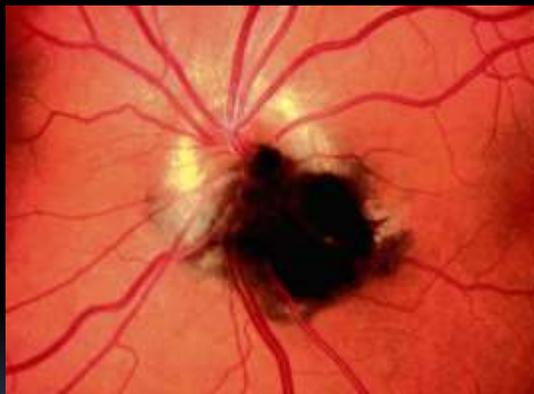
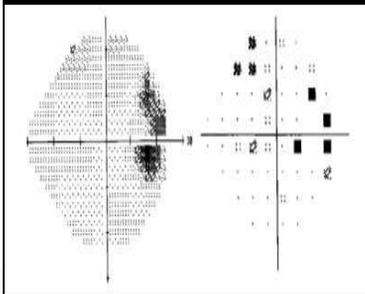
juxtapapillary Melanocytoma.

VF shows enlarged blind spot, due to tumor extension past the disc margin.



juxtapapillary Melanocytoma

Melanocytoma



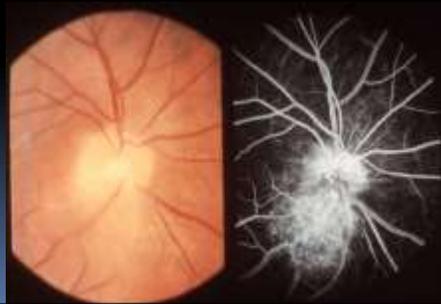
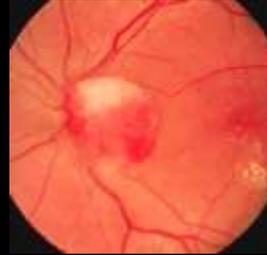
Melanocytoma cause nerve fiber bundle defects that result from compression at the optic disc.

Capillary Hemangioma of the Disc

elevated, reddish
mass

obscuring the optic
nerve head

Dilated, tortuous
feeder vessels
arising from the
optic disc.



OPTIC NERVE Angioma

benign vascular
neoplasm

von Hippel-
Lindau syndrome

Multiple
angiomas

globular reddish
with a dilated
feeding artery
and a tortuous
draining vein

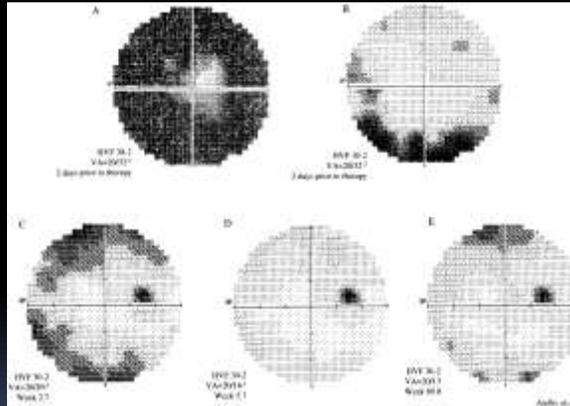


Optic nerve Angioma

VF: generalized
constriction

VF : before therapy
baseline visit

VF : Follow up after
antivascular
endothelial growth
factor (AVEGF)



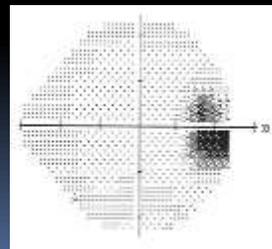
Astrocytic Hamartoma

Benign glial cell tumor

Multilobulated mulberry lesion



Visual field shows isolated
enlargement of the blind spot



Toxic/ Nutritional Optic Neuropathy

Drugs :

ethambutol, amiodarone,
immunosuppressive
medications as methotrexate
and cyclosporine

Eating and social habits

drinking, smoking

Toxic Optic Neuropathy

Ethambutol toxicity

5 weeks after onset of
visual symptoms.

Bilateral optic nerves
temporal pallor

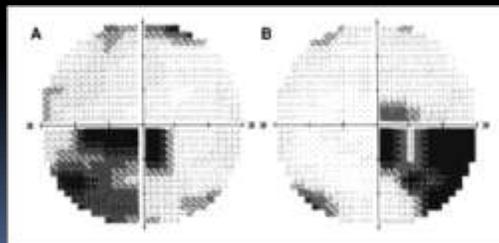
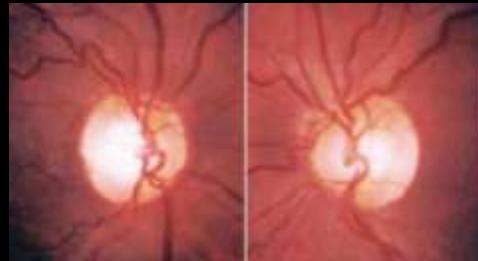
VF

3 months after onset of
visual symptoms.

**Bilateral central scotoma
inferior temporal quadrant
defects**

monthly VF till the medication is
discontinued.

If visual field defects are seen, stop
drug and follow-up fields every 1-3
months till VF have either improved
or stabilized



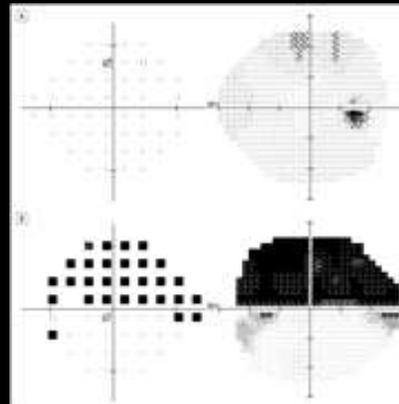
Anti-rheumatoid therapy

infliximab , Methotrexate

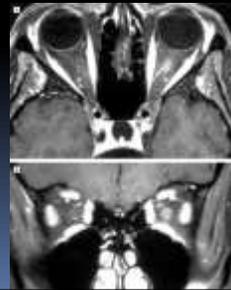
5-day history of decreased vision OS

OD normal VF

OS superior altitudinal defect.



MRI shows enhancement of the retrobulbar portion of the left optic nerve



Congenital Anomalies

Optic nerve hypoplasia

Optic pit

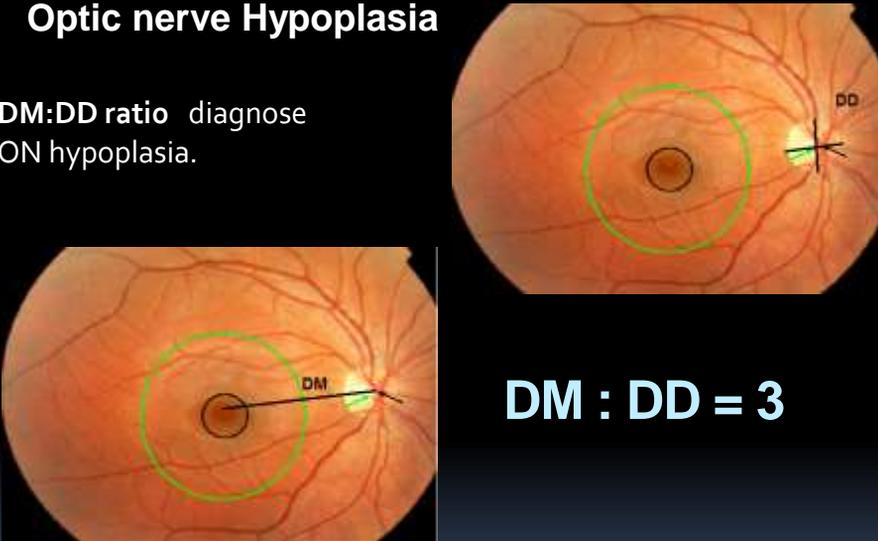
Optic disc drusen

Coloboma of optic nerve

Morning-glory syndrome

Optic nerve Hypoplasia

DM:DD ratio diagnose ON hypoplasia.



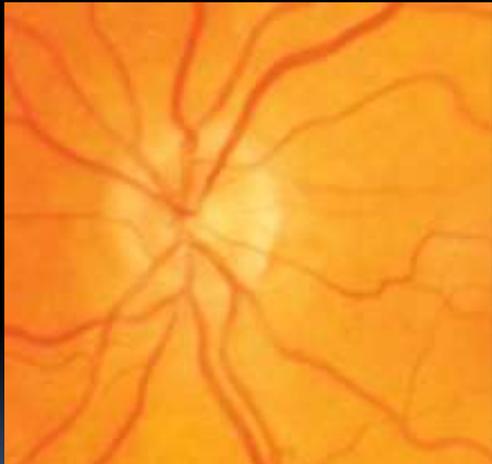
DM : DD = 3

$DM / DD > 3$ $DM/DD > 4$	ONH is suspected Optic Nerve Hypoplasia
------------------------------	--

Optic nerve hypoplasia

Small ONH due to a low number of axons.

double ring sign



Septo-optic dysplasia

OPTIC NERVE HEAD HYPOPLASIA

All components are small

small disc area
small cup
small neural rim

38
32
44
61

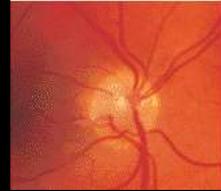
45
68
54
54

Superior Optic Nerve Hypoplasia Topless disc

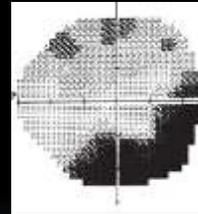
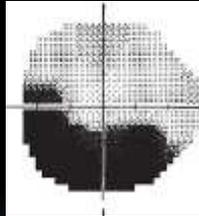
Segmental ON hypoplasia

Superior Optic Nerve Hypoplasia

Superior entrance
of CRV

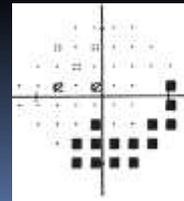
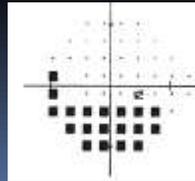


Pallor of superior
disc



Superior
Peripapillary halo.

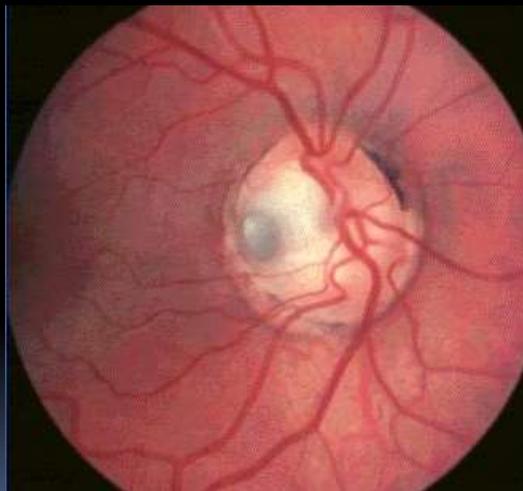
Superior nerve fiber
layer is absent



VF: inferior arcuate defects
consistent with superior
segmental optic nerve
Hypoplasia

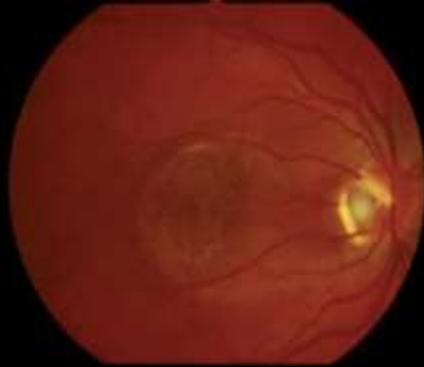
Congenital pit of the optic nerve

grayish circular
pit along the
temporal aspect
of the optic cup.

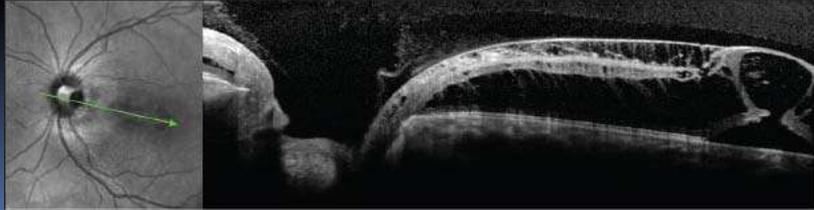


OPTIC PIT

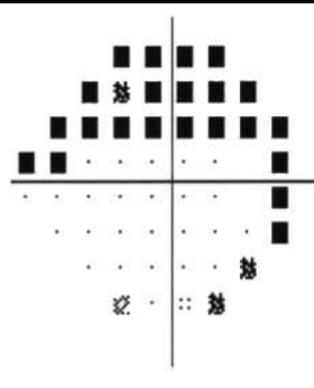
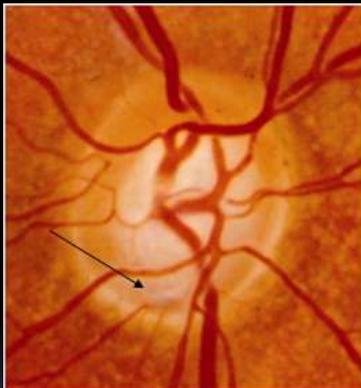
Associated with serous sensory detachment of the macula.



Schisis-like cavity extending from the optic disc to the macula & CME



Optic pit

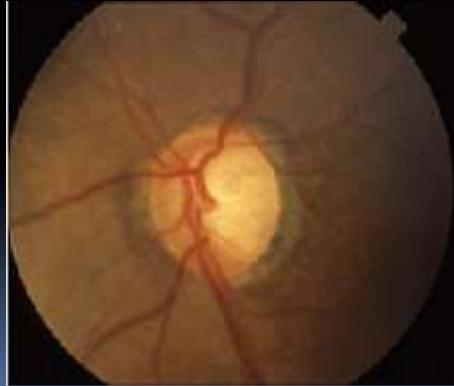


Corresponding upper field defect

Acquired pit of the optic nerve

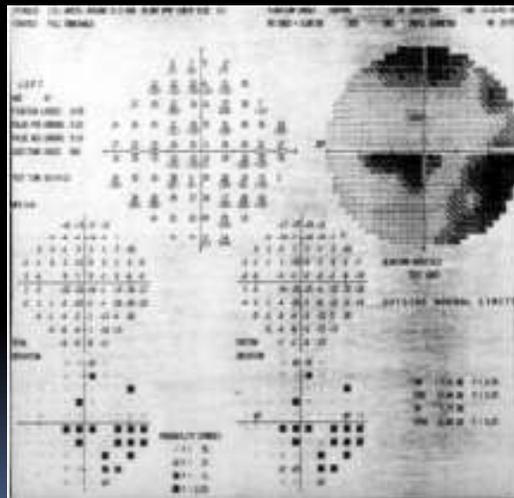
Acquired pit of the optic nerve. Note circular depression in the superior half of the optic cup.

Disk photo of APON
visual field of OD
from an NTG patient



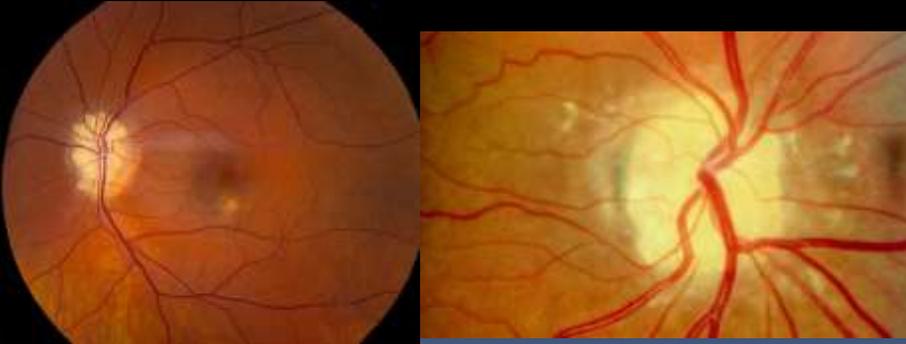
Visual field of acquired optic pit

dense inferior nasal step and paracentral scotoma which corresponds to location of superior APON in the optic nerve.



optic disc drusen

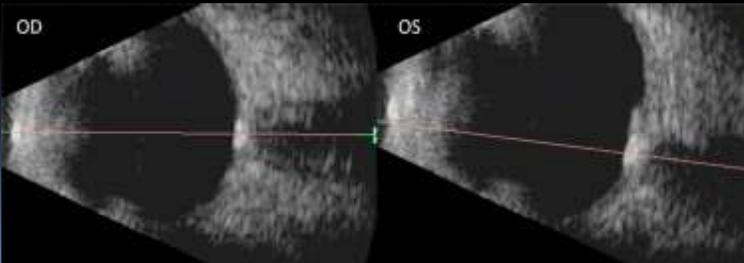
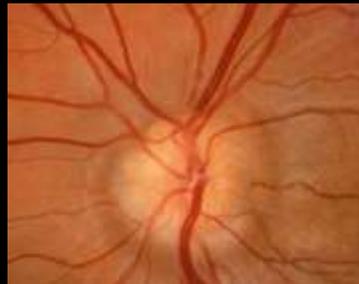
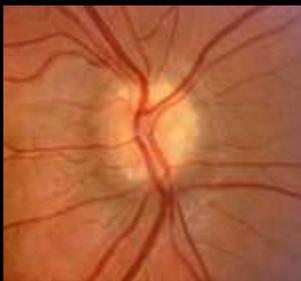
bilaterally elevated optic discs
with irregular scalloped margins
absence of central cup



ONHD

Numerous round, yellowish elevations in ONH, OD

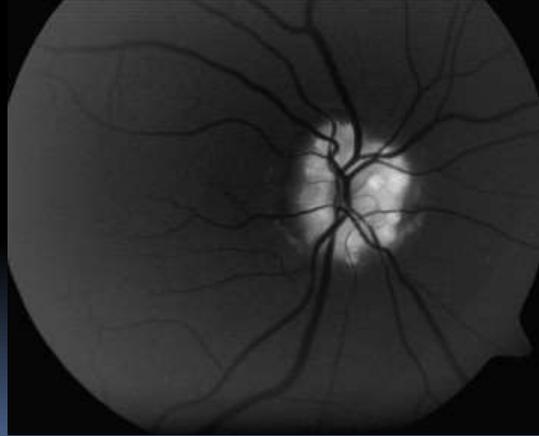
"lumpy-bumpy" yellow elevations within ONH, OS



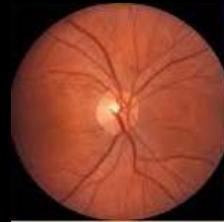
Autofluorescence in ONHD

Natural emission of light by biological tissues.

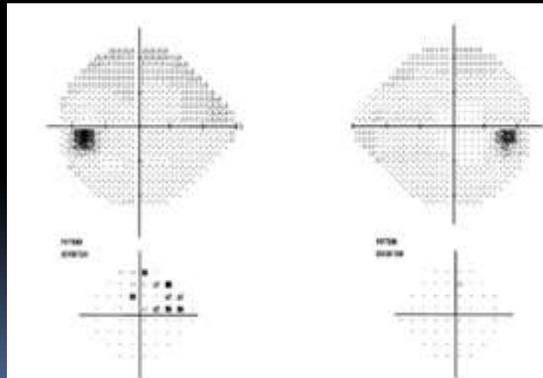
late staining of the drusen is due to the localized pressure of the drusen disrupting the blood tissue barrier in the optic nerve.



OD : normal

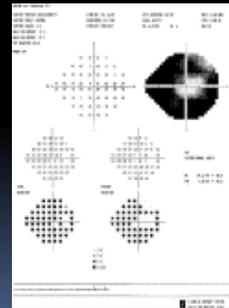
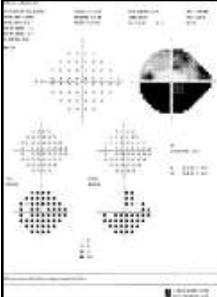
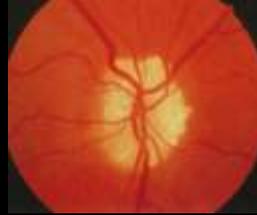
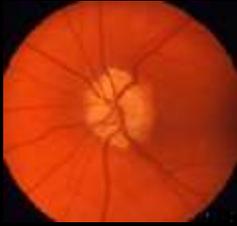


(OS) shallow superior arcuate scotoma



This scotoma represent early damage to the optic nerve from buried optic nerve drusen

superficial ONHD and peripapillary atrophy



OS :inferior altitudinal and superior nasal defects

OD : severely constricted field loss

optic nerves gliosis.
Small optic cup
no visible drusen.

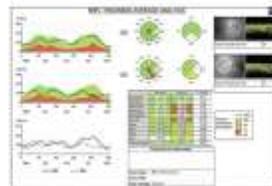
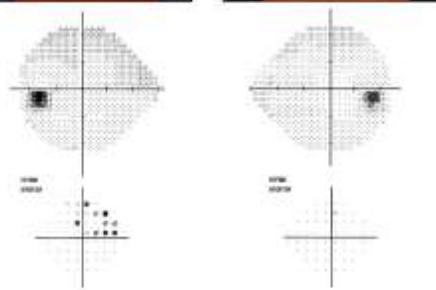
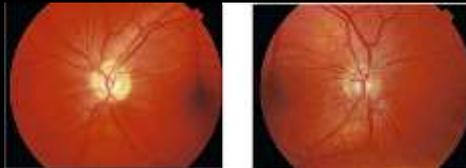
bilateral buried ON
drusen, confirmed by
ophthalmic ultrasound

Right normal VF.

Left superior arcuate
scotoma

Left OCT RNFL shows a
abnormal sector inferiorly
corresponds to the
superior arcuate scotoma.

This scotoma and RNFL
defect may represent early
damage to the optic nerve
from buried optic nerve
drusen (OND).



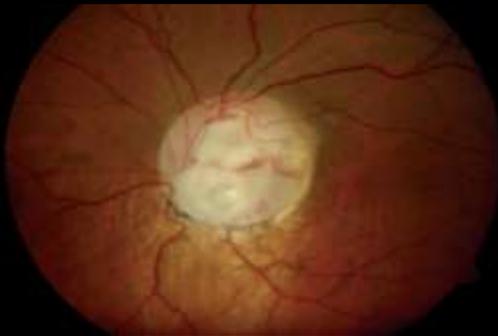
optic disc drusen

No TTT for complications of ONHD.

Follow up by serial VF, OCT nerve fiber layer , and fundus photography.

Lower IOP to prevent further nerve fiber layer and optic nerve damage

Coloboma of optic nerve head



incomplete closure of the embryonic fissure.
white, bowl-shaped excavation of the inferior ONH.



Inferonasal coloboma of ONH,retina & choroid.

Optic disc & choroidal coloboma

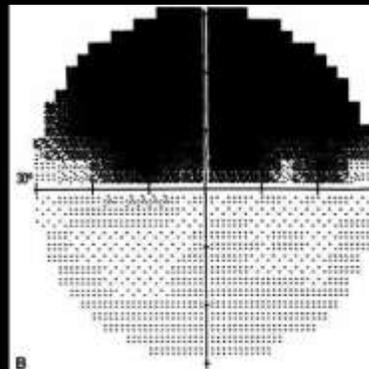
OS : choroidal coloboma inferior to the optic disc .

OCT shows that the sclera is bowed posteriorly

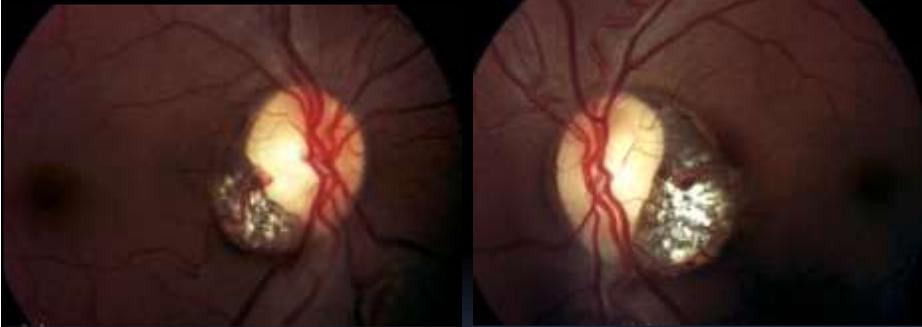


The optic disc is excavated because of the accompanying disc coloboma

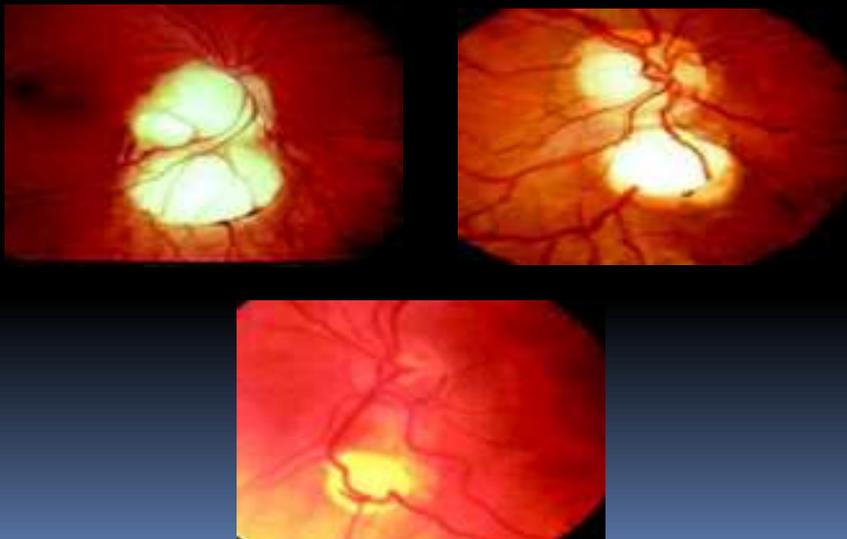
Optic disc coloboma and visual field deficit mimic glaucoma



Bilateral coloboma of optic nerve



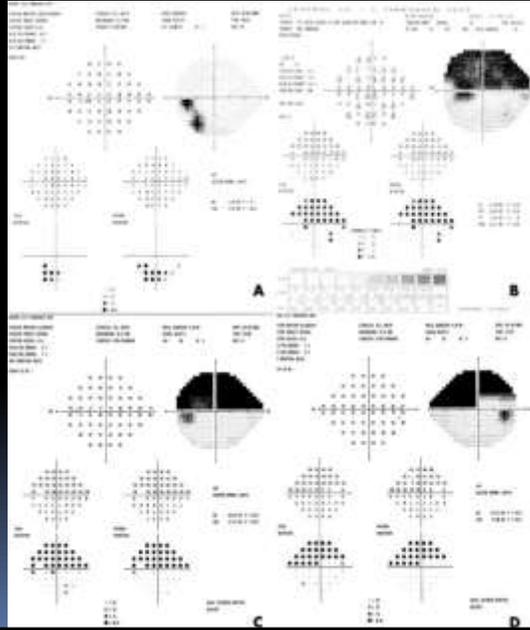
Optic disc duplication or coloboma



Humphrey visual fields

Double blind spot

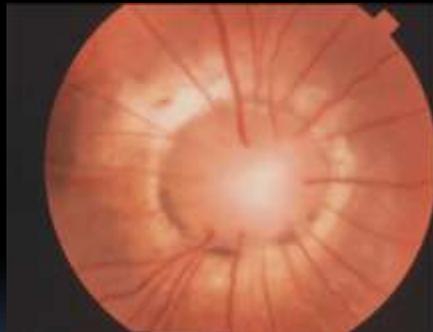
Superior hemifield defects



Morning Glory syndrome

Funnel-shaped excavation
Of the posterior
pole

surrounded by an
elevated
annulus of
Chorioretinal pigment .



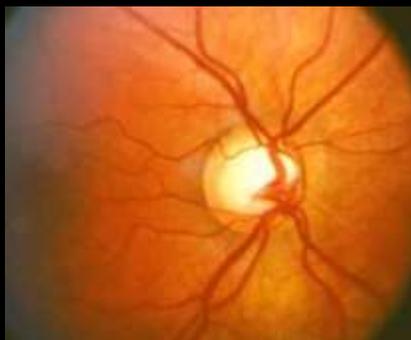
Megalo-optic disk

congenital anomaly,
visual acuity 20/20 in each eye

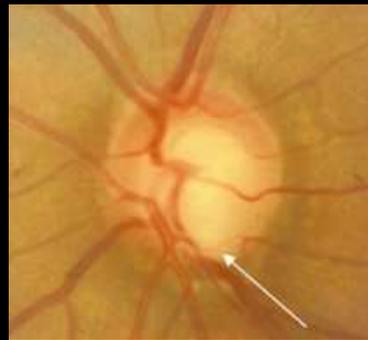
OS : normal disk



DD glaucoma
DD morning glory



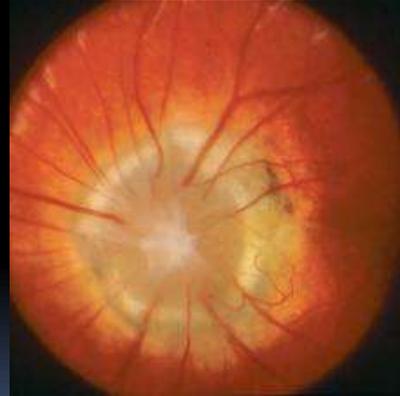
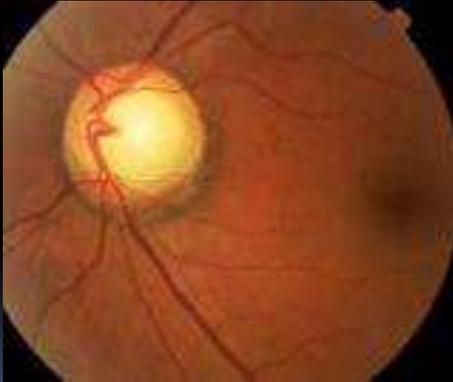
Megalopapilla



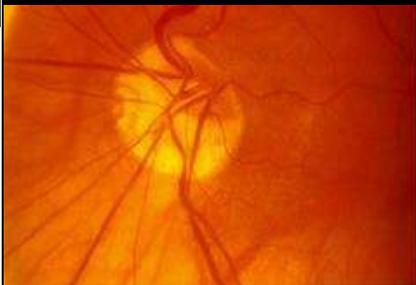
inferior notch

- 1-normal visual field
- 2- normal daily IOP curve.
- 3- The cupping is symmetric vertically and horizontally
- 4- No focal notching of the rim

MEGALOPAPILLA AND MORNING GLORY SYNDROME



Tilted Optic Nerve disc



bilateral

optic nerve exits the eye at an oblique angle.

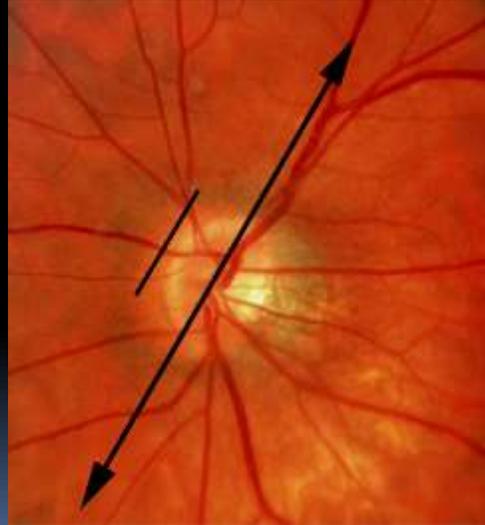
The blood vessels enter the globe at an oblique angle.

superiortemporal disc is raised, simulating disc swelling,

inferiornasal disc is flat or depressed .

Tilted Optic Disc

- oval-shaped disc with the long axis at an oblique angle.
- situs inversus of BV
- peripapillary atrophy
- Inferonasal Chorioretinal thinning and pigment accumulation
- posterior staphyloma



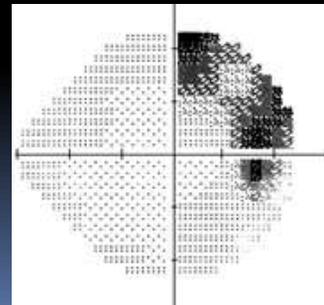
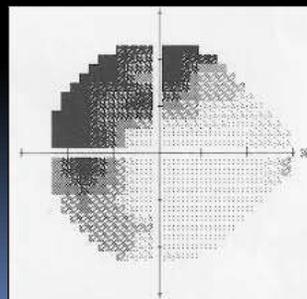
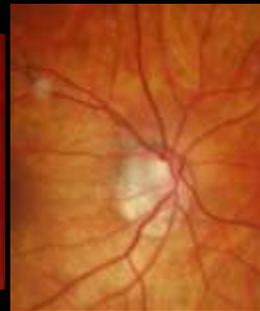
Bilateral tilted disc syndrome

VF show :

**bitemporal upper
quadrantanopia,**

mimic chiasmal lesion

**it crosses the
midline –
diagnosis of
tilted disc
syndrome.**



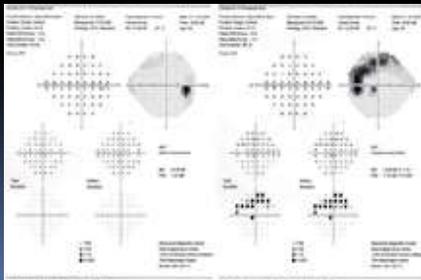
Tilted Disc



Right normal optic disc

left optic disc larger
Temporal pigmentary changes
atrophic infero-temporal regions

OD: normal



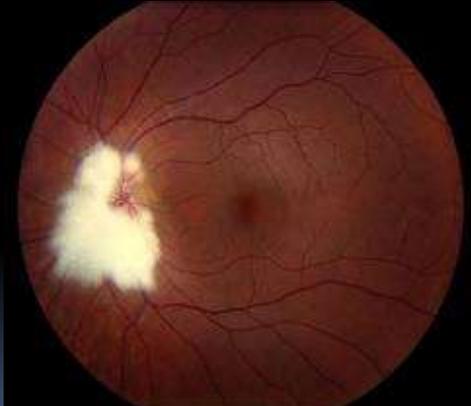
OS : superior arcuate scotoma

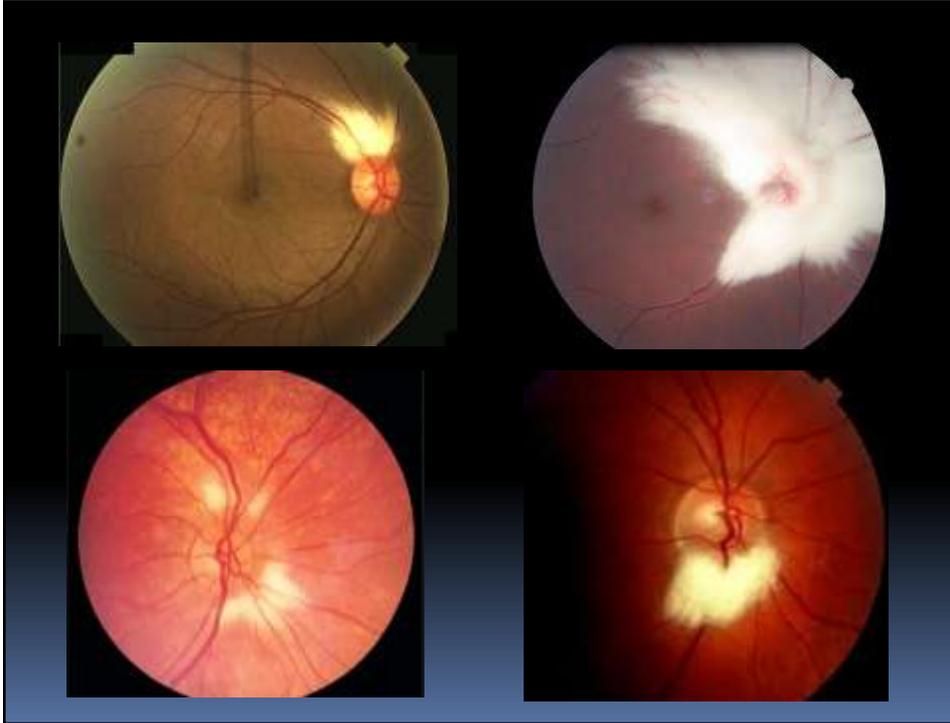
Myelinated nerve fiber layer

white, feathery patches follow the NFL bundles striated appearance

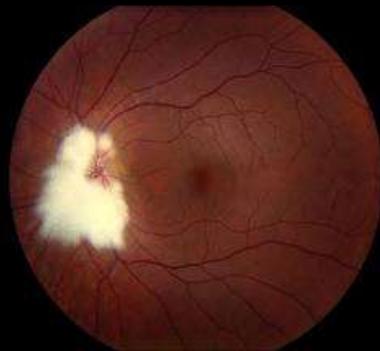
Normally myelination of the optic nerve fibers stops when it reaches the optic disc (shortly after birth).

1% myelination extending a variable distance into the retina.





Myelinated optic nerve fibers



enlarged blind spot
corresponding to the
area of myelination.

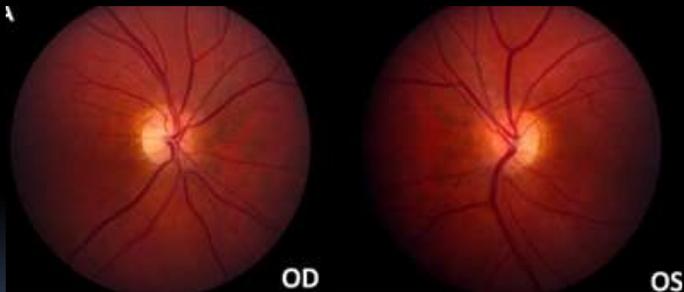
Hereditary optic neuropathy

Dominant optic atrophy (DOA)

Mitochondrial diseases

men and women alike have a 50% chance of transmitting the mutation to each child.

Death of RGCs

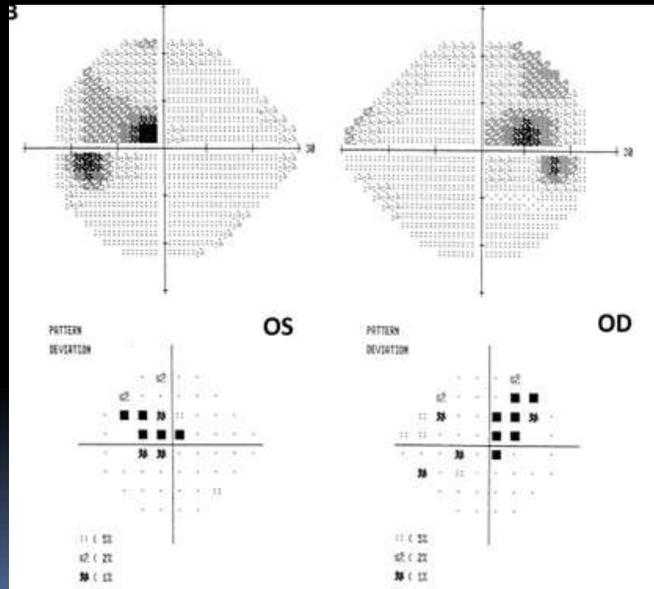


Bilateral temporal optic disk pallor

Humphrey VF

bilateral
cecocentral
scotoma

bilateral super
temporal field
defect



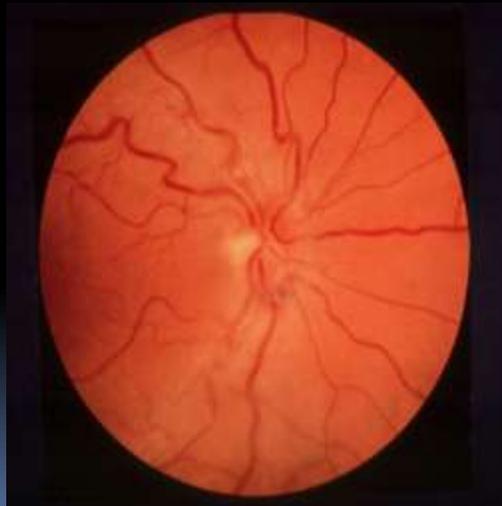
Leber's hereditary optic neuropathy

Mitochondrial inherited

women transmit the
mutation to all children,
men do not

degeneration of retinal
(RGCs), their axons

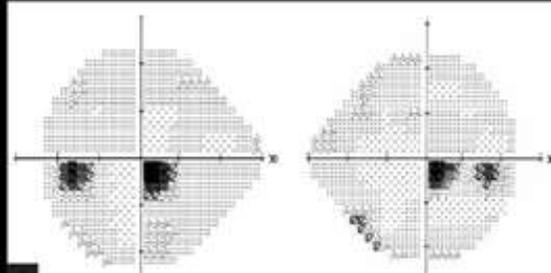
- . Disc hyperemia
- . dilated capillaries
- . Tortuous BV
- . Swelling of peripapillary
nerve fiber
- . Bilateral optic atrophy



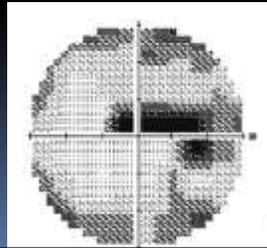
Leber's hereditary optic neuropathy (LHON)

acute loss
of central vision

Subsequent
bilateral optic
atrophy



Central or
cecocentral
scotoma are the
classic VF defects



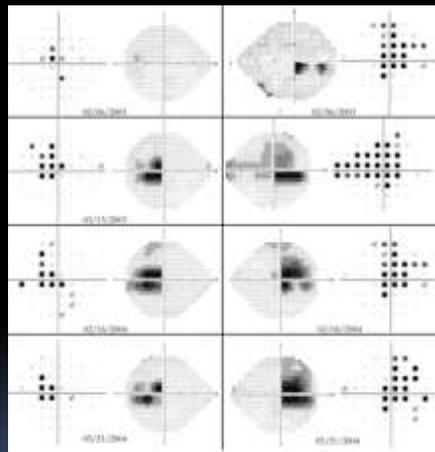
Leber's hereditary optic neuropathy

normal VA at
baseline in the fellow eye

VF of normal eye shows
subclinical findings

worse rapidly over weeks to
months to similar cecocentral
scotoma

**Quantitative VF is helpful
to know the natural history of
LHON and in understanding the
pathology and pathophysiology
of this disease ,
timing of gene therapy**



COMPRESSIVE OPTIC NEUROPATHY

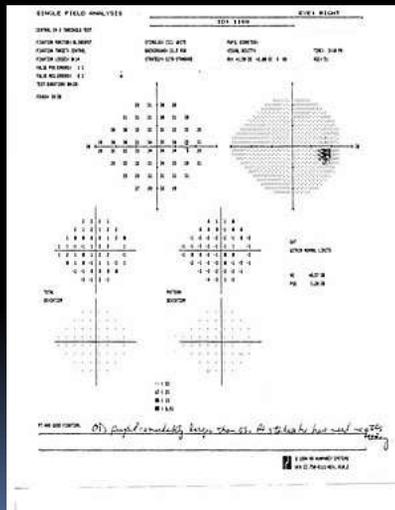
Thyroid Eye Disease (Graves' ophthalmopathy)

Autoimmune disease

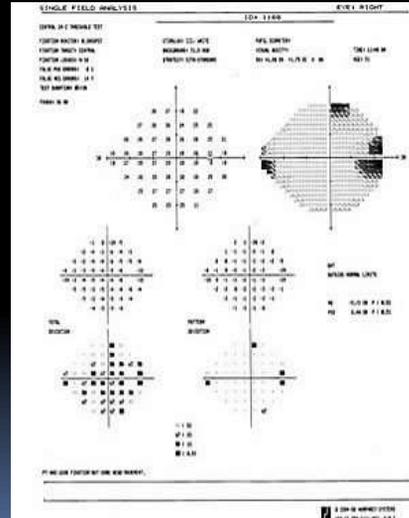
Bilateral proptosis
diffuse enlargement
of recti m.



VF: after Thyroidectomy



VF: under Medications



Take home message

1. Visual field test is critical in the diagnosis and follow up of optic neuropathy.
2. Visual field defects in optic neuropathies take several patterns including central, diffuse, arcuate, and altitudinal defect.
3. The pattern of visual field defect is not specific of any etiology and any type of field defect can occur with any optic neuropathy.
4. altitudinal defects are more common in ischemic optic neuropathies
5. central, or cecocentral defects frequently accompany toxic/nutritional and hereditary optic neuropathies.

