

Papilloedema is it or isn't it

PAPILLOEDEMA vs PSEUDOPAPILLOEDEMA



Why is papilloedema important to me?

An interesting fact:

30% of brain tumours present with visual problems

- ◆ Brain tumours can cause papilloedema
- ◆ Brain tumours are very rare
- ◆ Most papilloedema is not caused by a brain tumour

PAPILLOEDEMA

Papilloedema is bilateral optic disc swelling that is secondary to elevated intra-cranial pressure (ICP). Can be unilateral. May be asymmetric.

All patients presenting with papilloedema should be suspected of having an intra-cranial mass until there is proof to the contrary. Not a Primary condition.

Incidence is 2.5 per 100,000 (1,2)

Papilloedema - causes

Anything which leads to raised I C P

Most common cause (80%) is Idiopathic Intracranial Hypertension (I I H)

- Primarily a disorder of women of child-bearing age
- Higher than average BMI is also a risk factor

Papilloedema optic nerve function

SIGNS & SYMPTOMS (1, 2)

Visual Acuity

In its early & acute stage it is unusual to have visual problems

Transient Visual Obscurations – Where the vision goes grey/disappears. ‘dimming out’

Thought to be due to ischaemic effect of mechanical pressure on ONH blood supply

Papilloedema optic nerve function

Pupils

No RAPD present until end stage

Colour Vision

Not affected in the early stages

Diplopia

Associated CN VI can cause horizontal diplopia

Papilloedema

Optic Nerve Head Appearance

Blurred disc margins

Absence of physiological cup

Vessels Obscuration- Swollen/Elevated RNFL

Flame haemorrhages

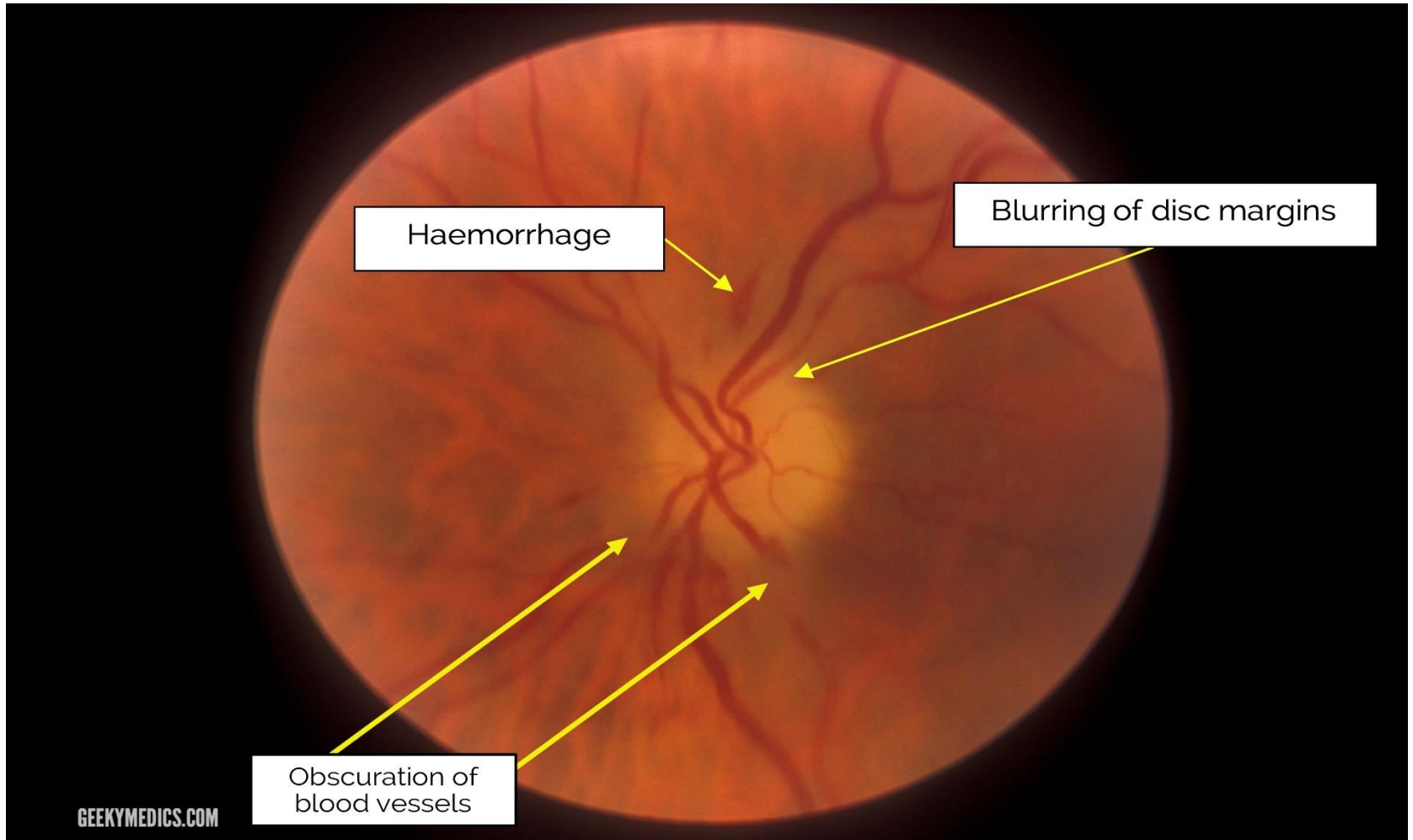
Absence of spontaneous venous pulsation (SVP)

SVP- Present or Absent (SVP + or SVP -) - present in 80-90% of normal population.

Presence of SVP is reassuring.

Absence of SVP is not diagnostic.

OBSCURATION OF THE BLOOD VESSELS SECONDARY TO RNFL ELEVATION



Papilloedema optic Nerve function

Visual Fields

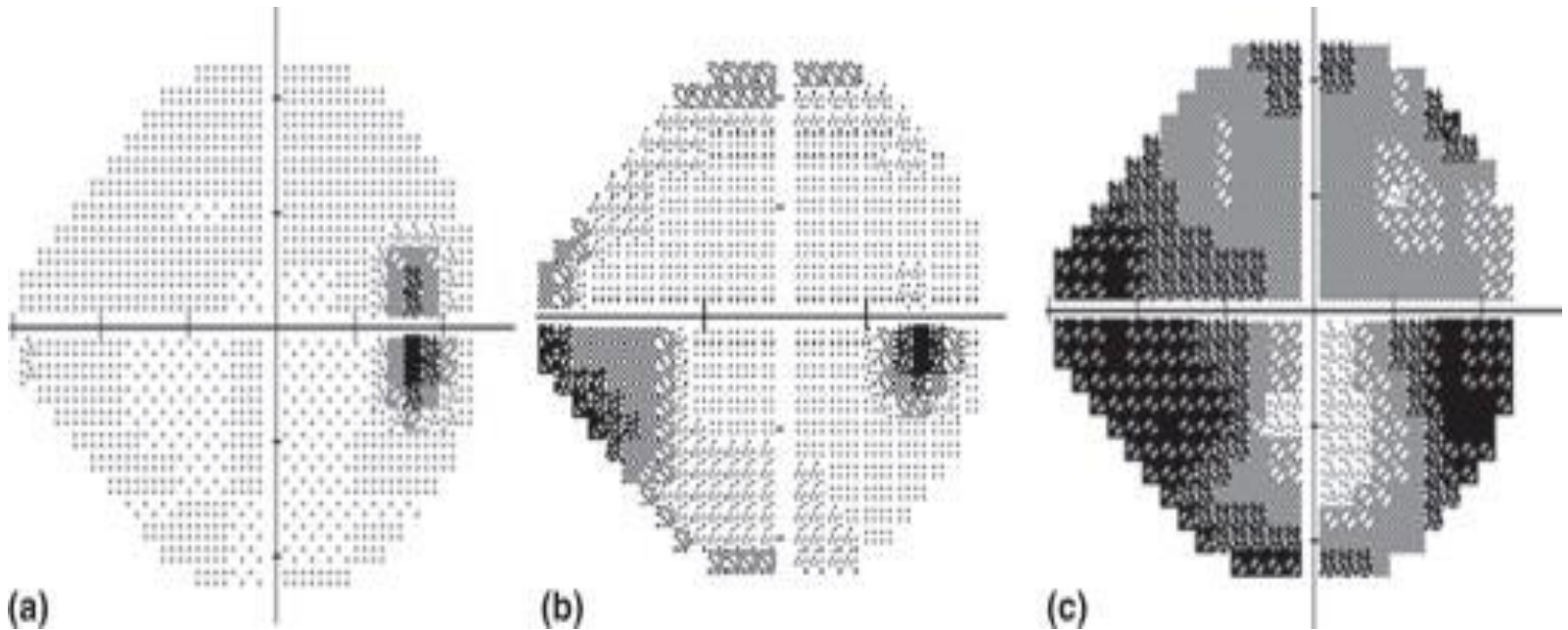
Enlargement of the Blind Spot, may only occur in later stages

Followed by Inferior VF changes due to Superior Pole damage

VF deteriorates as the condition progresses

VF mostly useful as monitoring response to treatment

Progressive Visual Field Changes in Papilloedema



Papilloedema

OCT Findings in True Papilloedema

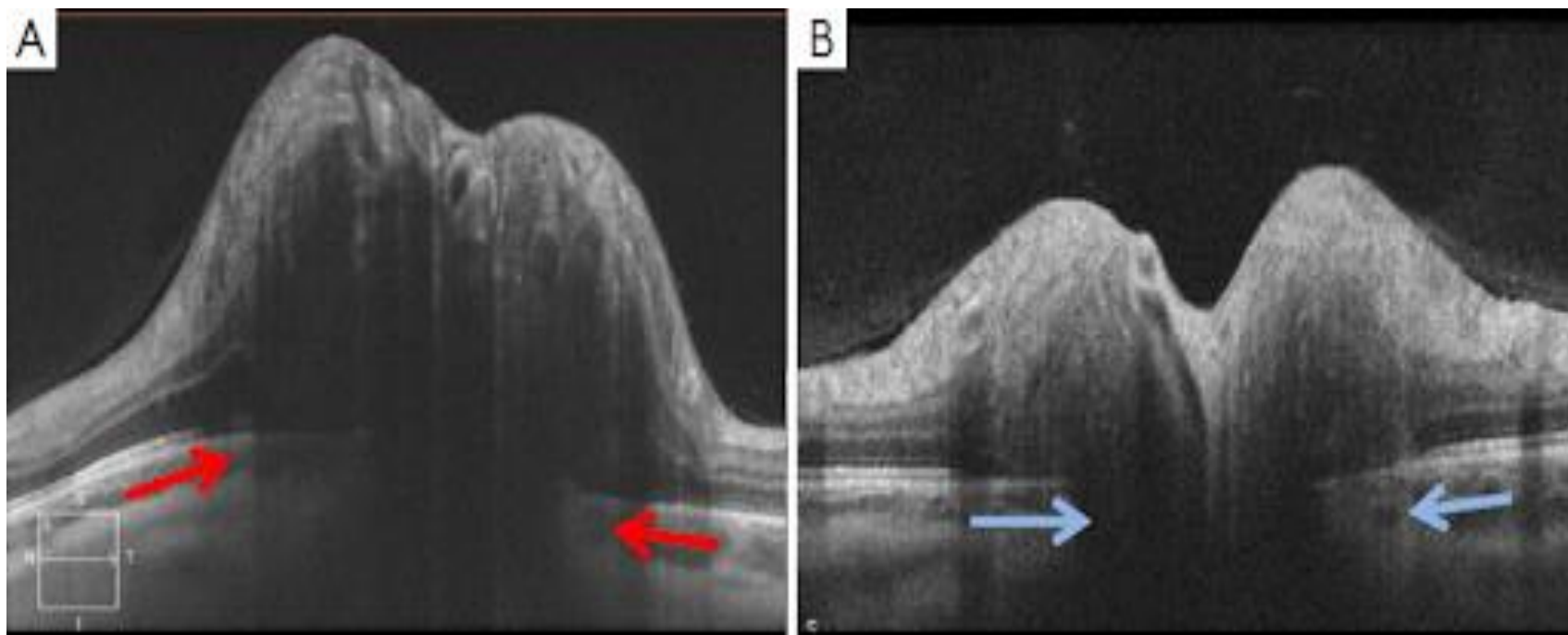
ONH Volume especially Central Thickness

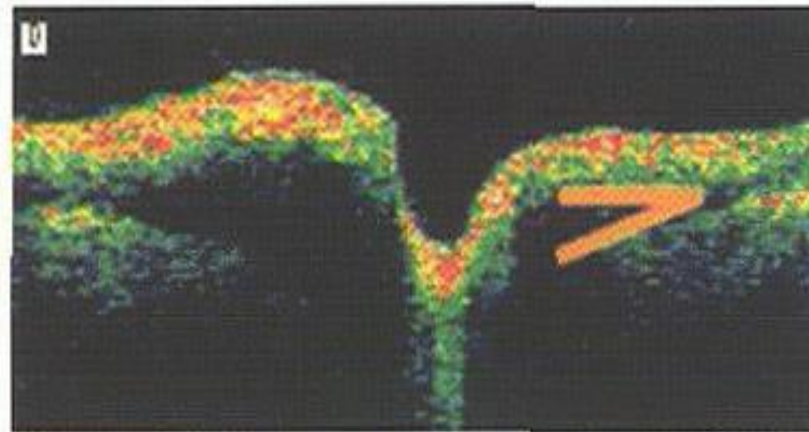
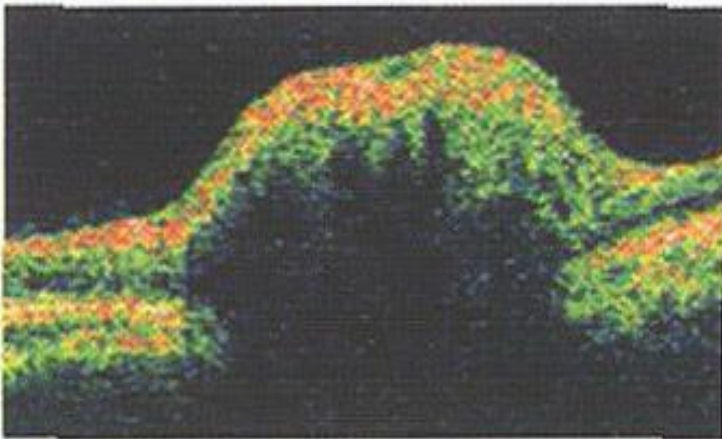
Peripapillary RNFL Thickness- Especially Spectral Domain

Area of sub-retinal hyporeflective space

Angle between the RNFL and the Optic Nerve. 'Lazy V' Sign

Forward Bowing of Bruch's Membrane/RPE seen with Enhanced Depth Imaging. INWARD vs DOWNWARD angulation

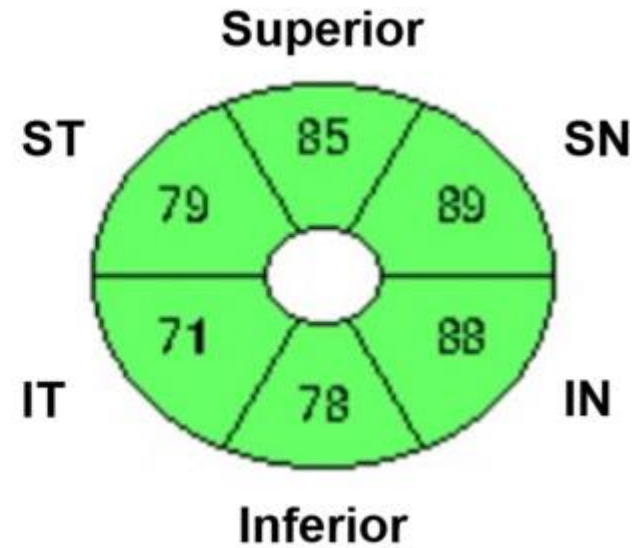
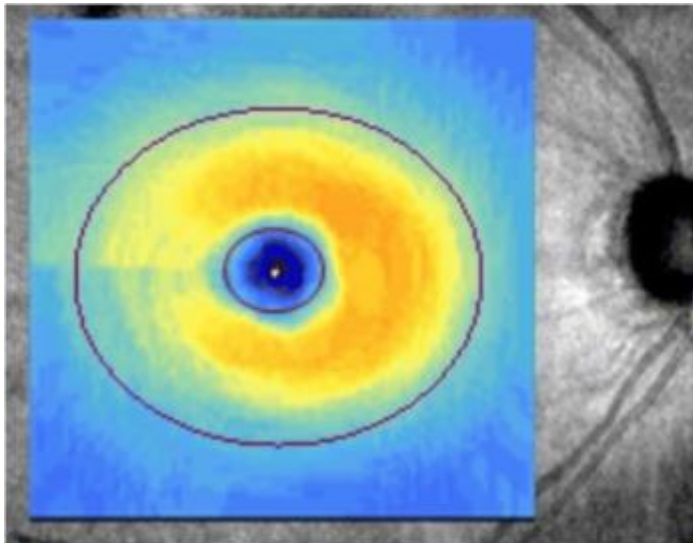




12. In a patient with visible optic disc drusen, disc topography (left) shows an elevated optic nerve head with a lumpy internal contour and abrupt end to the hyporeflective space. Compare this with the smooth internal contour and V-shaped hyporeflective space between the retinal pigment epithelial and photoreceptor layers in a patient with optic disc edema (right).

Image courtesy: Johnson LN, et al.
Differentiating optic disc edema from
optic nerve head drusen on OCT.
Arch Ophthalmol. 2009;127(1):45-9.

MACULAR GANGLION CELL-INNER PLEXIFORM



Poll Number 1.

OCT findings indicative of Papilloedema would be all of these apart from

- a) Increased RNFL peri-papillary thickness
- b) Area of sub-retinal hyper reflective space
- c) Lazy-V Sign
- d) Forward-bowing of Bruchs Membrane/RPE

Papilloedema

Symptoms

Headaches

Pulsatile Tinnitus/ 'Whooshing' Sound ('Ringing')

Nausea & Vomiting – usually 1st thing in the morning

Enough to wake you from your sleep

Worse upon physical exertion coughing/sneezing

Worse when moving from seated to standing

Papilloedema

Transient Visual Obscurations (TVOs)

An Episodic loss of vision.

‘Greying Out’ or ‘Dimming’ of the vision

Associated with eye movement and body position changes

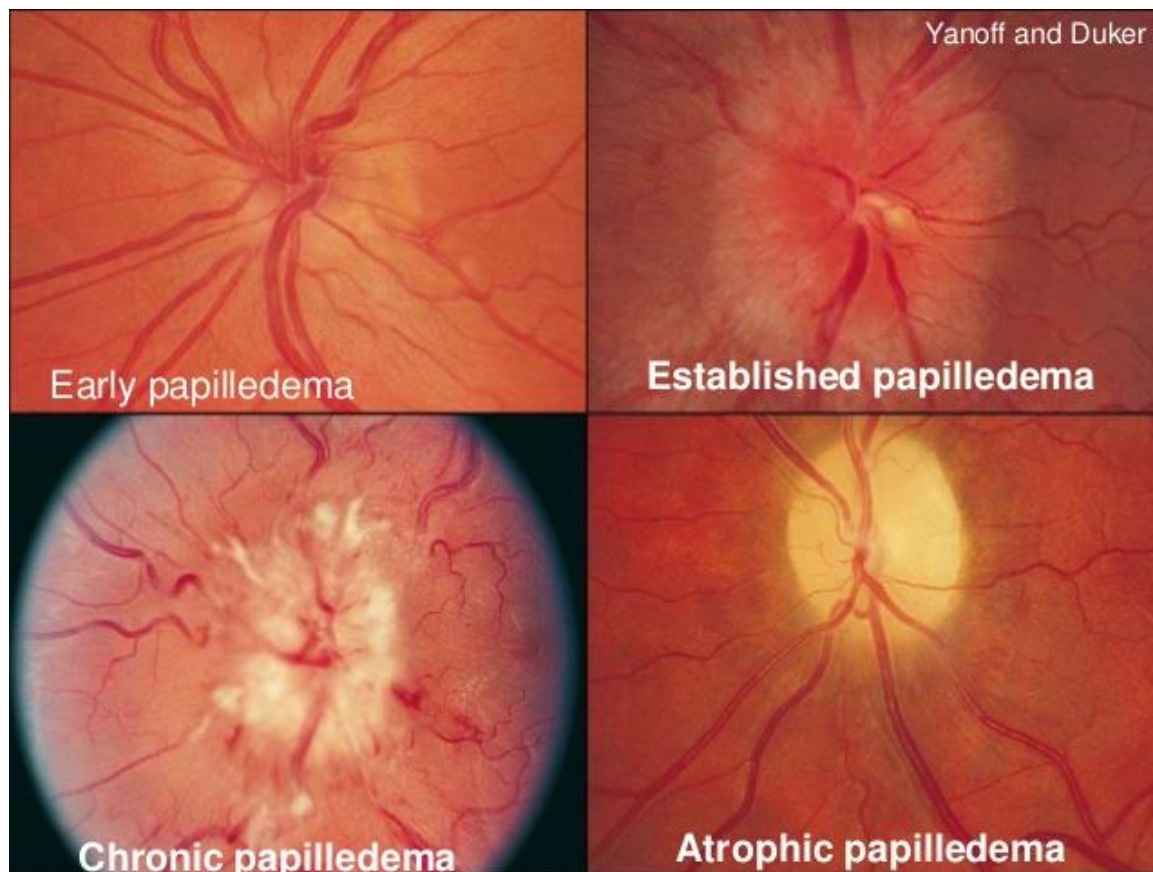
May last 1-2 secs- Full Monocular Field Loss

Poll Number 2

The typical headache characteristics associated with a papilloedema include all of these apart from

- a) Projectile vomiting mainly in the morning
- b) Worse with Valsalva manoeuvres
- c) Strong enough to wake you from your sleep
- d) Worse when you stand up

EARLY, ESTABLISHED, CHRONIC, ATROPHIC IMAGES



Papilloedema

Papilloedema 4 Stages

Early

Established

Chronic

Atrophic

Early Stage

Visual Symptoms are absent

Mild Disc Hyperaemia

Indistinct/Blurred Disc Margins-Elevated (Nasal)

Tortuous Vessels, possibly small haemorrhages

'C-Shaped Halo'

Absence of SVP

C-Shaped Halo, Temporal Quadrant unaffected

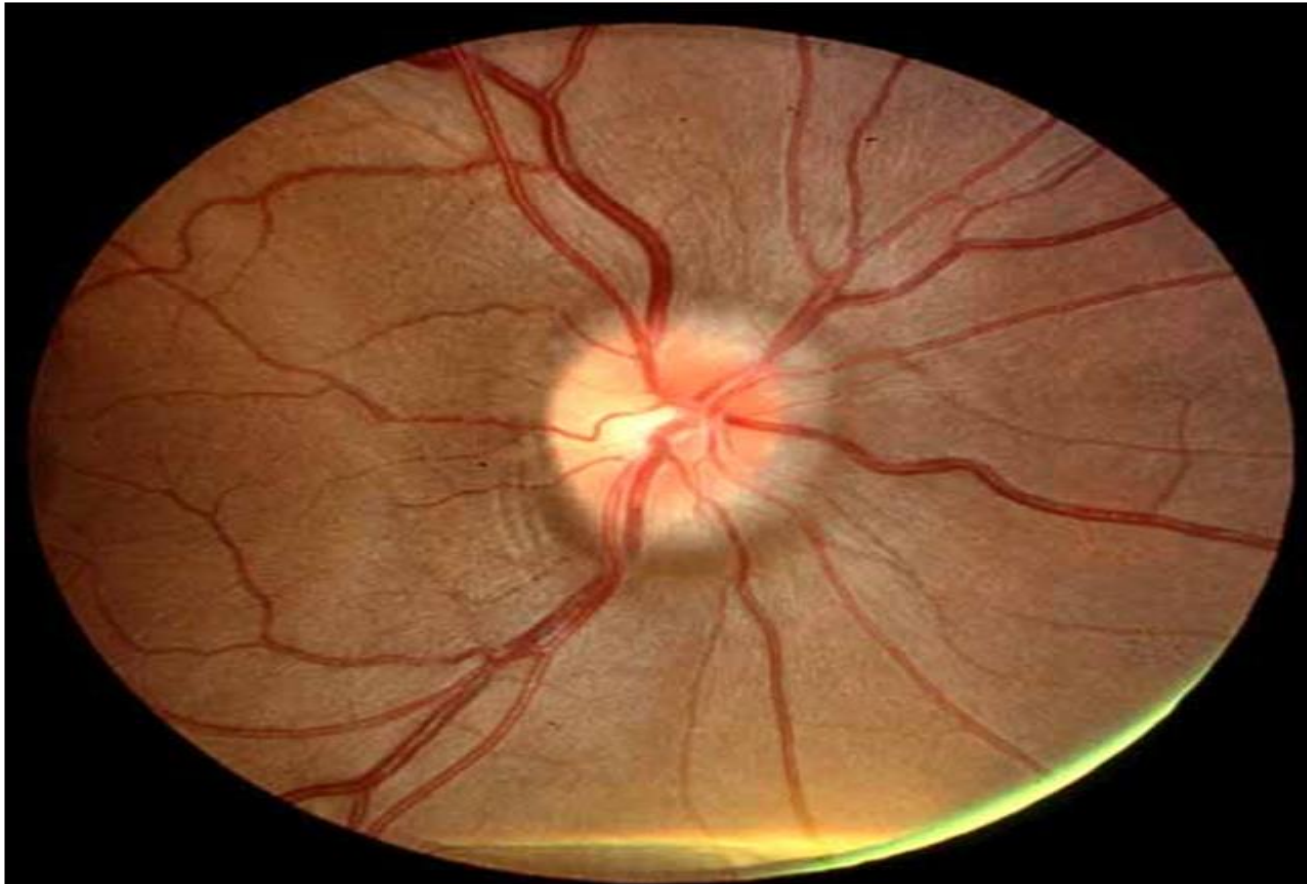


Figure 6. Grade I papilledema is characterized by a C-Shaped halo with a temporal gap.

Papilloedema

Established

Visual symptoms are absent

Transient Visual Obscuration

Cup Filled Obliteration- filled up with exudates

Severe Disc Hyperaemia

Venous engorgement, peripapillary flame haemorrhages

Cotton Wool Spots

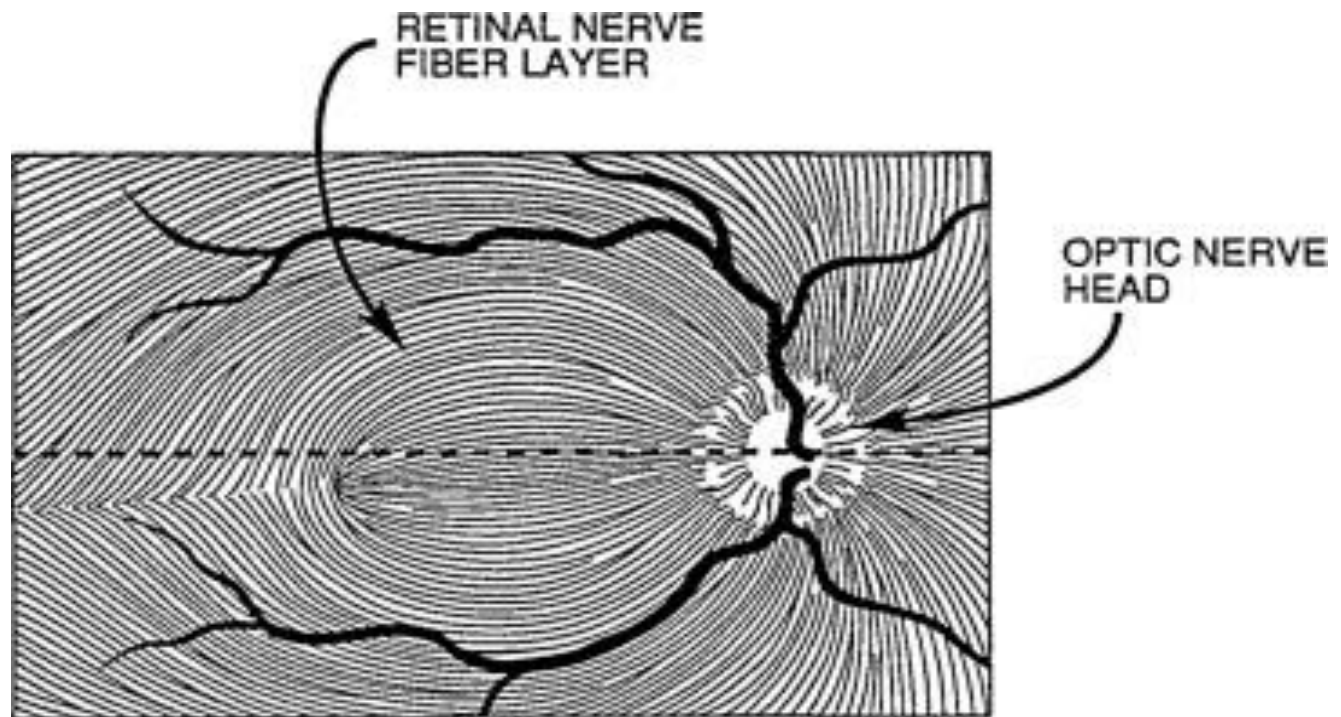
Circumferential Retinal Folds (Paton Lines)

Hard exudates radiate outward



Figure 6. Grade I papilledema is characterized by a C-Shaped halo with a temporal gap.

NORMAL RNFL DISTRIBUTION- RADIAL OR VERTICAL





Paton's Line

Circumferential retinal folds due to papilledema

Papilloedema

Chronic

Visual Acuities are variable

Visual Fields show constriction

Optic disc shows like a 'Champagne Cork' Appearance

Pale Disc

CWS & and Haemorrhages are absent

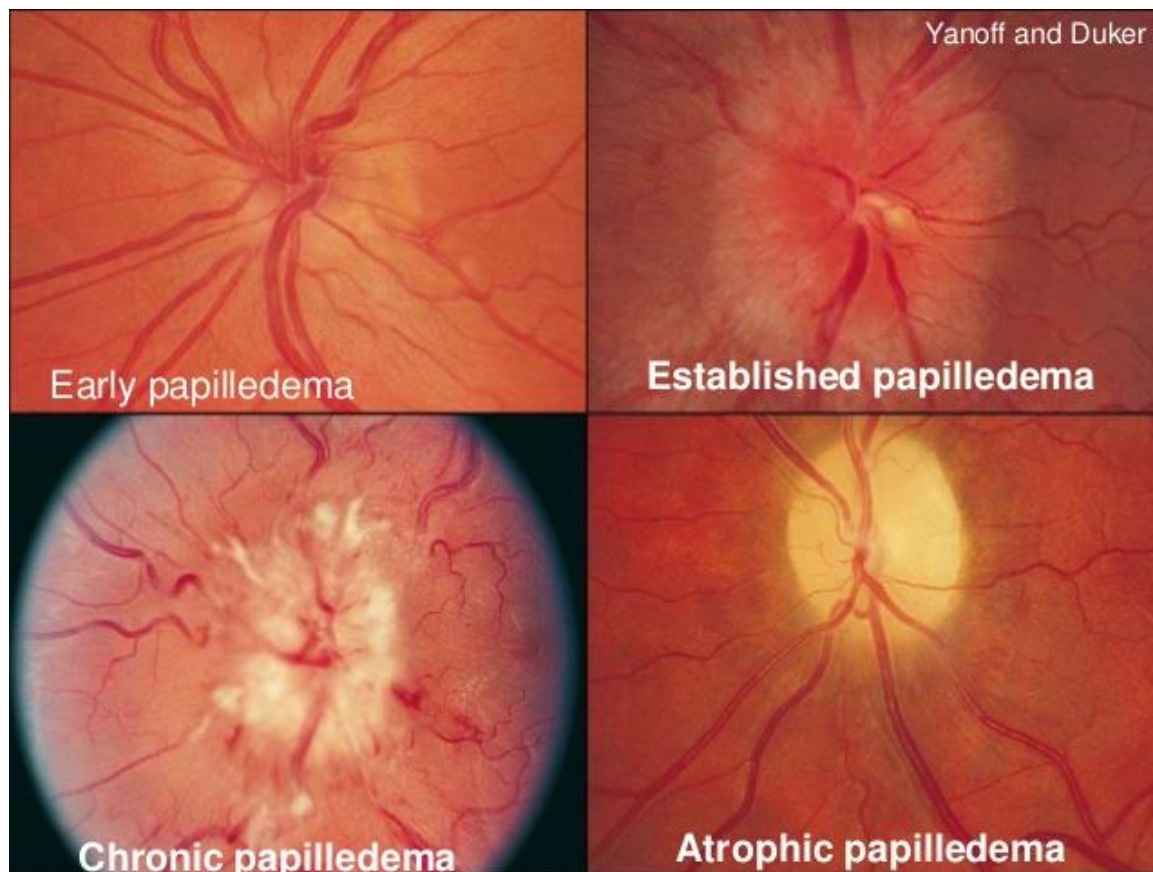
Atrophic

Flat Disc, indistinct Margins

Visual Fields Compromised

Visual Acuity is severely impaired

EARLY, ESTABLISHED, CHRONIC, ATROPHIC IMAGES



Papilloedema-obligatory signs

Retinal Changes associated with Papilloedema

Macular changes = Oedema-Sub Retinal fluid

Macular changes = lipid hard exudates = "Macular Star"

Macular haemorrhages

CWS & Exudates

'Paton's Line'- Chorio-retinal Folds around the disc, circumferential in nature

Papilloedema

SVP as a diagnostic sign in Papilloedema (3)

Present in 80%, absent in 20% of normals

Was it recorded as being present previously

However SVP +, or a SVP – can be VERY helpful to the ophthalmologist who is triaging referral –

Moorfields Data (5) Ms Sui Wong Study on I I H & SVP- to check the validity of dictum 'presence of SVP excludes raised ICP'.

13 of 106 had high ICP, 11 of 13 had SVP (86%)

SVP

- Spontaneous pulsation of veins in or around the disc
- Caused by differential between IOP and ICP
- Some clinicians even advocate mild pressure on the globe through the upper eyelid
- Best seen via DIRECT OPHTHALMOSCOPY as 15x mag

HOSPITAL TESTS

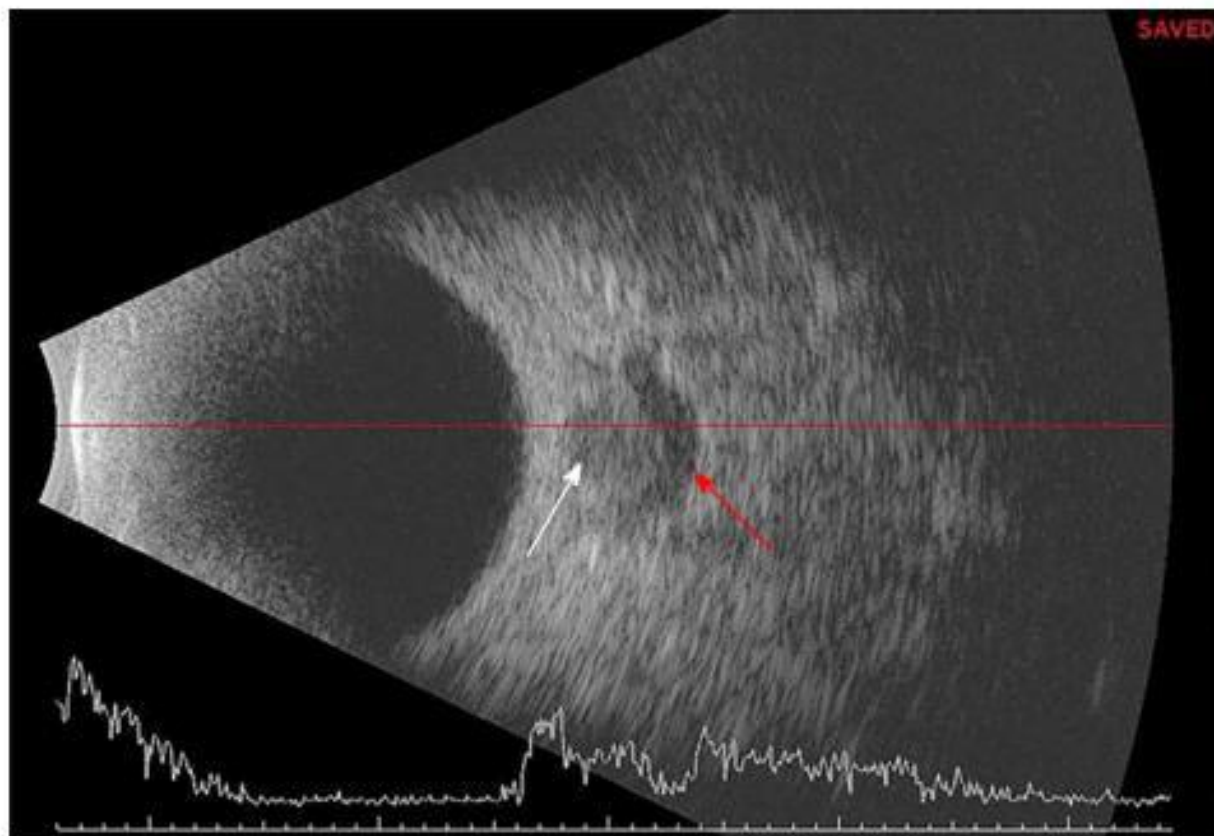
Ultrasound B Scan looking for Crescent shape (92% success rate in detecting Papilloedema) (4)

CT of the head to rule out Space Occupying Lesion
CT Venogram – Checking Venous Sinuses for Thrombosis

Fluorescein Angiography – Papilloedema leaks

OCT RNFL thickness can be used for Baseline data
OCT Bruchs membrane – looking for forward bowing BM/RPE

Ultrasound b crescent sign



Poll 3

Which of the following Hospital screening tests would not be performed to screen for Papilloedema

- a) Ultrasound A & B
- b) CT scan & CT Scan Venogram
- c) Macular Pigment Density Test
- d) OCT ONH Analysis

Papilloedema

Moorfields data from Emergency Dept. James Acheson

Over a 6 month period there were 61 children who were referred for suspicious optic disc appearance.

3 had papilloedema

29 were asymptomatic

So optometrists do tend to have a high rate of false positive referrals

DOPS 2019 Study found community optometrist 21% false diagnosis of papilloedema from fundus photography in comparison to hospital optometrists 7% false diagnosis (6)

DOPSS Study

‘Fundus Photography in isolation is highly sensitive but poorly specific for papilloedema detection.’

‘Using this method alone to screen has the potential for harm as over diagnosis occurs’

PSEUDOPAPILLOEDEMA

No set protocols exist for the differentiation of pseudopapilloedema & papilloedema

- Detailed history & symptoms
Sore Neck, feeling dizzy, trouble walking
- Where available - multi-modal non-invasive techniques: digital retinal photo ,OCT & visual fields.

Pseudopapilloedema

Differential Diagnosis

Optic Neuritis

Anterior Ischaemic Optic Neuropathy (AION)

Optic Nerve Head Drusen (OHND)

Tilted Optic Disc

Congenital Crowded Disc

Unilateral causes

Key Features	Optic Neuritis	AION	Papilloedema
<u>LATERALITY</u>	Unilateral	Unilateral	Bilateral** (can be asymmetrical)
ONSET	Sudden	Sudden	Insidious
LOSS OF VISION	Sudden	Sudden	Gradual if any
DISC SWELLING	None unless papillitis (uncommon)	Present	Marked
VISUAL FIELD DEFECT	No definitive pattern	Hemi-Altitudinal	Enlarged Blind Spot
PUPILS	RAPD +VE	RAPD +VE	RAPD - VE

Pseudopapilloedema

Optic Nerve Head Drusen

It is the most common cause of pseudopapilloedema accounting for 75% of cases (2)

Bilateral in 75%

Nasal > Temporal, Peripheral > Central Location

Congenital occurring approx. 1-3% of General population
Caucasians > Afro-Caribbean's

2 different types Visible & Hidden

Hidden- located beneath the disc surface and cause disc elevation,
become more visible in 2nd-3rd decade

May get Haemorrhages assoc. with ONHD

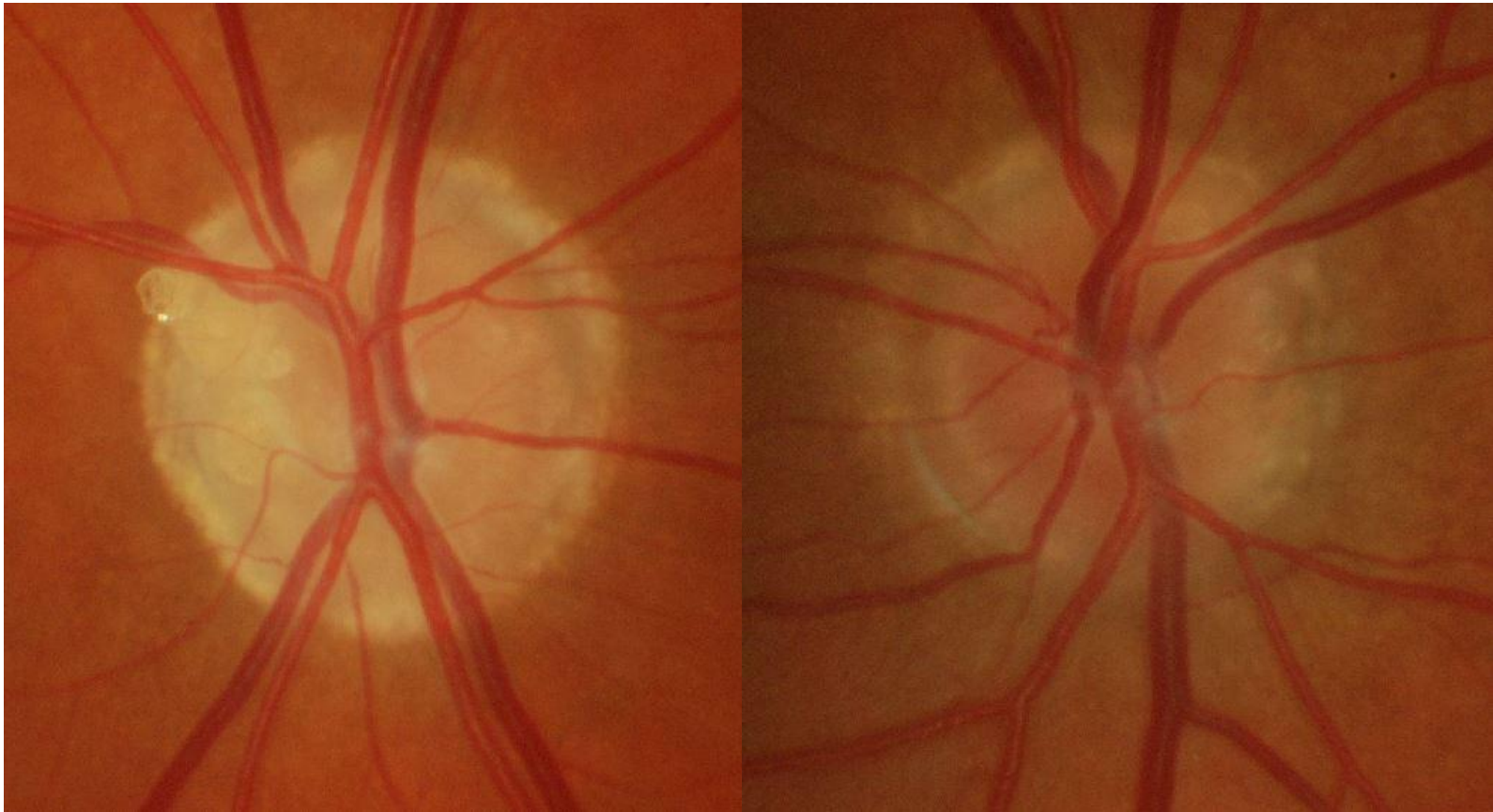
May get TVOs

May have VF defects similar To Glaucoma, respecting the
horizontal midline

Anomalous branching of blood vessels

Visible ONHD- cause anatomical/functional change RNFL

ONHD-drusenoid disc 'moth-eaten'



OPTIC NERVE HEAD DRUSEN ON ULTRASOUND B SCAN

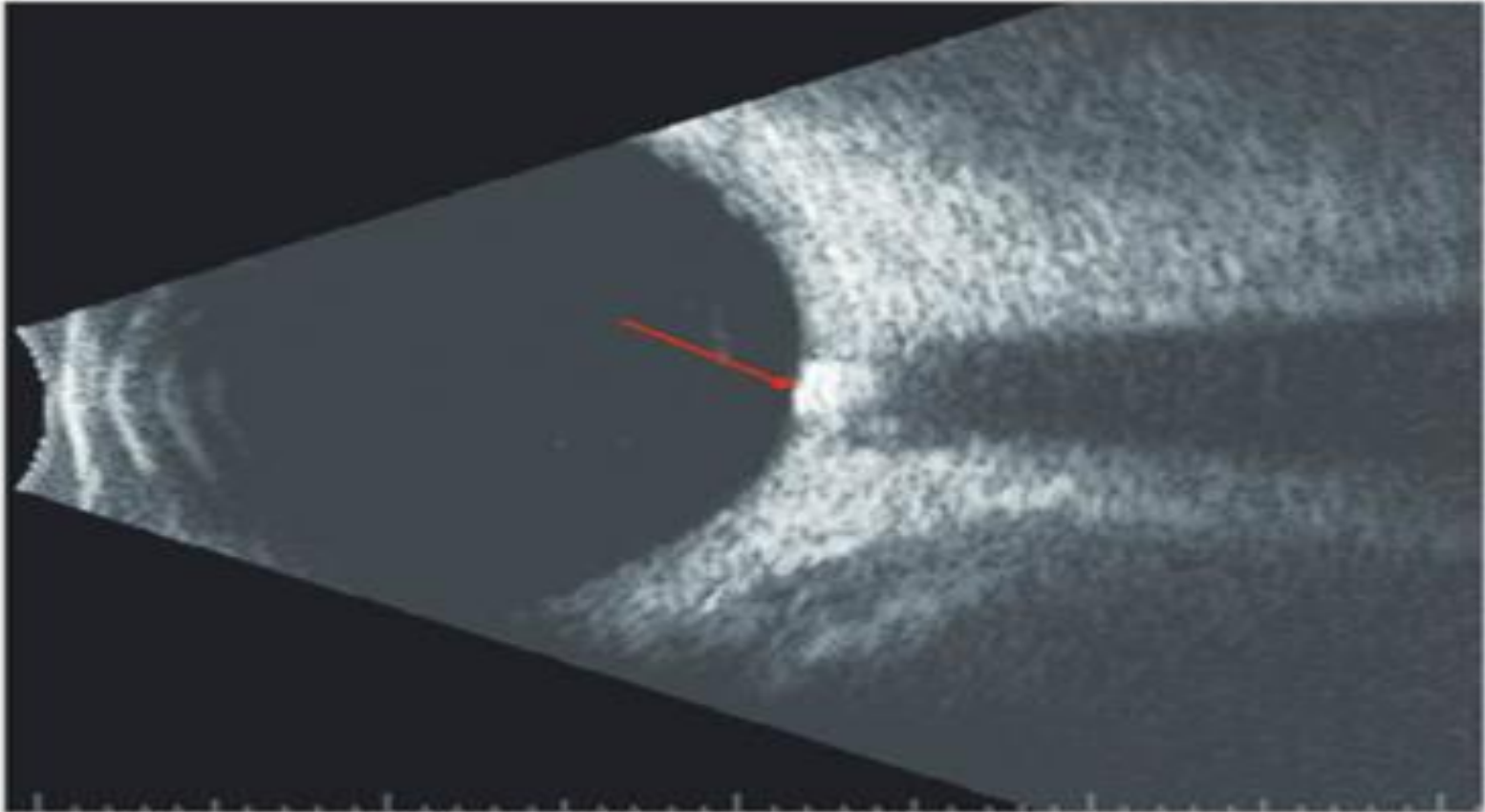


Fig. 4.15 Optic nerve head drusen on B-scan echography. The calcified drusen appear as an ovoid echogenic lesion (arrow) with a posterior acoustic shadowing.

Pseudopapilloedema

Tilted Optic Disc

0.4 to 3.5% prevalence, bilateral 37.5 to 80% of cases

Oblique Insertion of the nerve to the globe.

Nasal portion is elevated, temporal area depressed.

Nasal margin is blurred and gives a swollen like appearance

High assoc. with moderate myopic correction / astigmatism

Tilted discs



Pseudopapilloedema

Congenital Crowded Disc

Normal number of axons passing through a smaller posterior scleral foramen

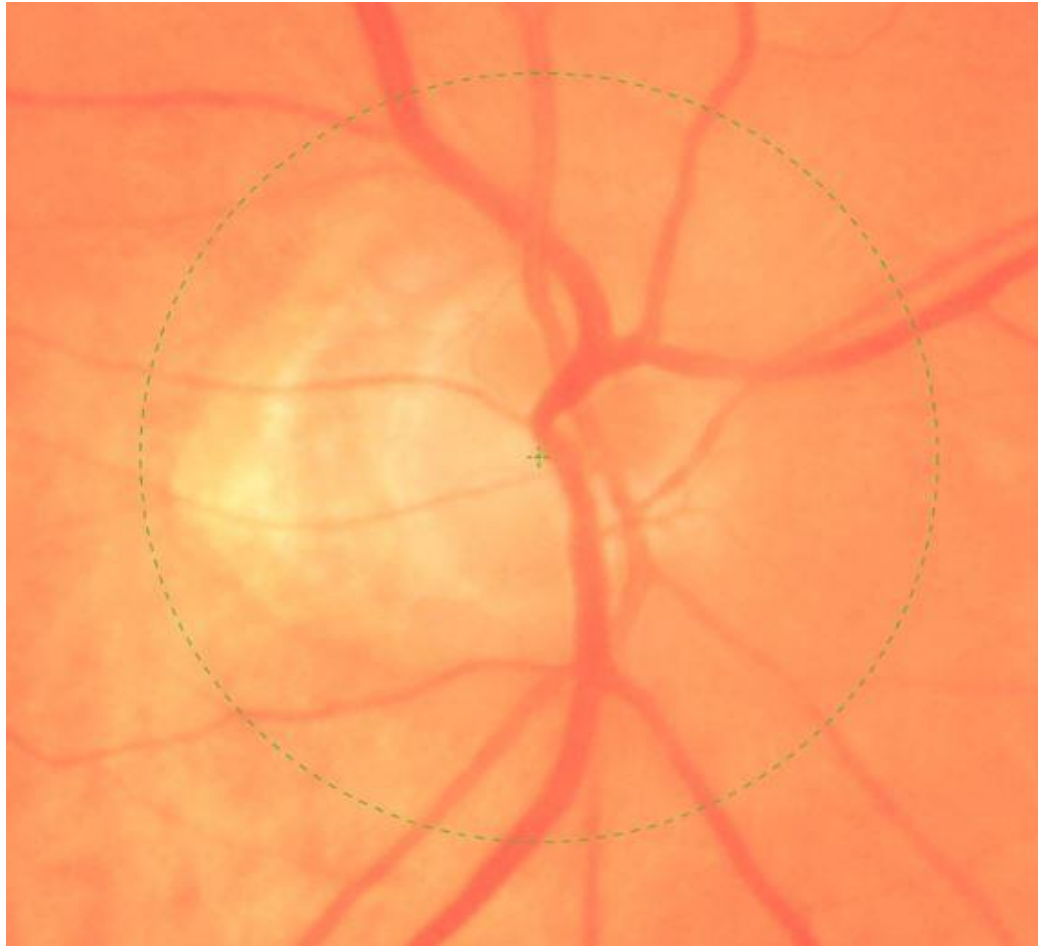
Appearance is a densely packed or crowded ONH.

Smaller than average disc, often assoc with hyperopic corrections
*** Measure disc as for glaucoma & record on referral***

Falsely hyperaemic in appearance, with no physiological cupping
superonasal & inferonasal blurred margins showing elevation

Sometimes loss of Physiological cup

Small, crowded disc looks hyperaemic



Differential diagnosis

Ultrasound B:

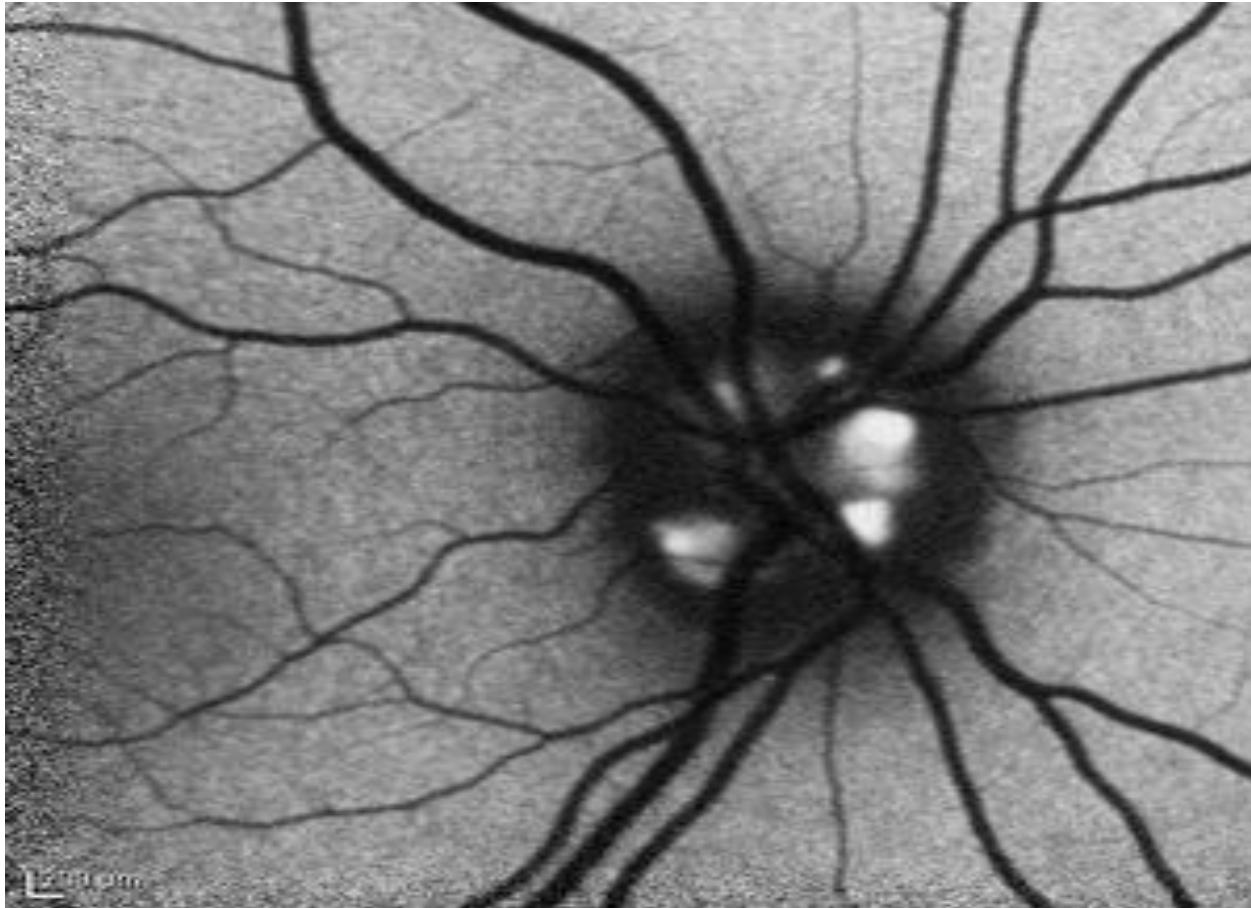
Normal optic nerve appears as a low reflectivity

ONH drusen appear as a hyper-reflective calcified bodies, like calcium in the bones it glows on scans

True papilloedema shows elevated optic disc with absence of hyper-reflective bodies

Papilloedema- Crescent Sign- 92% sensitivity in diagnosing
Papilloedema

Hyper-Reflective Bodies



Poll Number 4

Which of the following tests would not be used to detect Optic Nerve Head Drusen

- a) Ultrasound B
- b) OCT with Enhanced Depth Imaging
- c) Ultrasound A
- d) Fundus Auto Fluorescence

Recommendations

Recommendations

Fundus cameras- **Sequential photos** are very useful, if px is asymptomatic

The true power of an OCT is Repeatability

The best study/control for your patient is the patient

OCT Software Data base- Normative age related database is European Caucasians

Asians, Afro-Caribbean, high +/- Rx, or < 18yrs age

Sequential photos



OPTIC NERVE HEAD DISC DRUSEN DDX

Key Features	Optic Nerve Head Drusen	Papilloedema
Visual Symptoms	None	Transient visual obscurations
Visual Field	Various – can mimic glaucoma	Enlarged blind spot
Diplopia	None	Possible- CN VI
Headaches	None	Worse upon wakening
		Strong enough to wake
		Postural changes more
		Nausea/Vertigo
Optic Nerve Head	Elevation / pallor	Swelling/Hyperaemia
	Confined to Disc	Overlap disc margin
Retinal Features	None	Paton's Lines

PAPILLOEDEMA VS PSEUDOPAPILLOEDEMA

	Papilloedema	Pseudopapilloedema
Optic Nerve	Elevated	Elevated
Disc Colour	Hyperaemic	Normal
Disc Margins	Blurry	Sharp
Physiological Cup	Absent	Absent in Crowded Discs
SVP	Absent	Present in 80%
Haemorrhages	Peripapillary	Occasionally in ONHD
CWS & Exudates	Present	Absent

Differential Diagnosis

There is no ONE diagnostic tool that can confirm papilloedema.
Use all the weapons in your arsenal

However we do have in practice Multi Modal Non Invasive
Techniques to help us in our diagnosis.

Papilloedema is a condition which is usually diagnosed by
exclusion, rather than confirmation

Headaches – Does the patient have headaches that are worse when supine, but improves when they stand up? The headache is also worse when they cough, sneeze or bend down to tie shoelaces.

Pulsatile Tinnitus- ‘Whooshing’ Sound in their ears. ‘Ringing’, in time with their heartbeat.

Transient Visual Obscurations- are there ‘Greying Out’ or a ‘Dimming’ . Full Field Monocular loss. Lasts 1-2 secs

- Elevated Disc- If PX is asymptomatic take a picture and / or OCT and return 5-10 days later, if the images are identical, unlikely to be papilloedema
- DON'T PANIC
- Don't be afraid to call the on-call ophthalmologist for advice. NHS Service is available 24/7. On Call service
- 90% of Papilloedema will be Symptomatic, so only 10% will be Asymptomatic.

Importance of **H&S** is vital when distinguishing Papilloedema from Pseudopapilloedema. Characteristics of the pxs symptoms (postural/valsalva headache, visual obscurations, pulsatile 'whooshing' tinnitus) to guide urgency of referral.

True papilloedema unlikely to be picked up routinely in a completely **asymptomatic** px.

Importance of disc photos and disc OCT in conjunction with a detailed H&S in referral to HES for triage process.

Poll Number 5

Which of these is the only true real sign of Papilloedema?

- a) Absent Physiological Cup
- b) Disc Haemorrhages
- c) Vessel Obscuration
- d) Absence of SVP

Which of these is the only true sign of Papilloedema

- a) Absence of Physiological Cup- This can occur in a small crowded disc
- b) Disc Haemorrhages – this can occur with ONHD
- c) Vessel Obscuration- The most definite sign of Papilloedema
- d) Absence of SVP- ICP fluctuates throughout the day like IOP, so depending on when you see the patient

So the true sign to watch out for is Vessel Obscuration

Any further questions please do not hesitate to email

Lorcan.butler@thebraintumourcharity.org

For printable practice resources on signs & symptoms of brain tumours in children, please visit

www.headsmart.org.uk

For further information/donations, please visit

www.thebraintumourcharity.org

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