Optic Disc Swelling (including Papilloedema)

Background \[1,2\]

Optic disc swelling can be caused by a number of conditions including papilloedema. The term papilloedema refers specifically to optic disc swelling secondary to raised intracranial pressure (ICP). Patients with papilloedema usually present with signs or symptoms of raised ICP.

Not all optic disc swelling is papilloedema. Careful history and examination are needed to distinguish papilloedema from other causes of optic disc swelling such as central retinal artery or vein occlusion, congenital abnormalities and optic neuritis.

Optic disc swelling is distinct from optic disc atrophy which refers to a loss of nerve fibres at the optic nerve head and which results in a pale disc. See also separate Optic Atrophy article.

Causes of optic nerve swelling \[3\]

The most common causes of optic nerve swelling are non-arteritic anterior ischaemic optic neuropathy (35%), optic neuritis (31%) and intracranial pathology (14%).

- Unilateral optic disc swelling is more likely to be due to demyelinating optic neuritis, non-arteritic anterior ischaemic optic neuropathy, retinal vein occlusion and diabetic papillopathy.
- Bilateral swelling is more likely to be due to papilloedema, toxic optic neuropathy and malignant hypertension.

A fuller list of causes may be divided into:

Intracranial conditions

- Causes of raised ICP:
  - Tumour.
  - Cerebral trauma.
  - Intracerebral or subdural haemorrhage.
  - Cerebral inflammation/infection.
  - Cerebral abscess.
  - Idiopathic intracranial hypertension (pseudotumor cerebri), a condition with elevated CSF pressure and no mass lesion.
  - Respiratory failure.
  - Chiari malformation.
  - Acute mountain sickness and high-altitude cerebral oedema.
  - Lyme disease.
  - Some medications have been associated with raised ICP - eg, tetracycline, minocycline, lithium, isotretinoin, nalidixic acid and corticosteroids (both use and withdrawal).

Optic nerve conditions

- Optic neuritis.
- Optic neuropathy:
  - Arteritic ischaemic optic neuropathy (giant cell arteritis).
  - Non-arteritic anterior ischaemic optic neuropathy.
Toxic optic neuropathy (eg, methanol poisoning).
- Compressive optic neuropathy (eg, thyroid eye disease).

- **NB**: congenitally anomalous optic discs may appear swollen.
- Infiltration of the disc by sarcoid, glioma, lymphoma.

**Vascular causes**
- Retinal vein occlusion.
- Retinal artery occlusion.
- Malignant hypertension.
- Acute lymphocytic leukaemia (through infiltration of retinal vessels by immature lymphocytes).

**Conditions affecting the globe**
- Glaucoma: central vein occlusion.
- Long periods of weightlessness.
- Pan or posterior uveitis.
- Posterior scleritis.
- Irvine-Gass syndrome (see separate Macular Oedema article).
- Infiltration or inflammation (eg, sarcoid, leukaemia).
- Carbon dioxide retention.
- Parahypothyroidism.
- Uraemia.

**Assessment of optic nerve swelling**

Examination of the swollen optic disc pays close attention to signs which could support a diagnosis of papilloedema. If suspected this must be urgently confirmed or refuted by specialist examination. The separate article Examination of the Eye provides more detail on the method of fundoscopy and examination of optic nerve function.

**History**
- Full systemic, neurological and ophthalmic history.
- Specifically, assess for symptoms suggesting raised ICP. Ask about any headaches and their characteristics and associated symptoms (eg, nausea/vomiting, worse on waking, coughing and bending ± pulsatile tinnitus, all of which can indicate raised ICP). See also separate Raised Intracranial Pressure article.

**Examination of the disc**
- Before dilating the pupils assess for a relative afferent pupillary defect (RAPD) - see 'Evaluation of optic nerve function', below. This is a helpful sign in many conditions.
- Examine the margin of the disc: establish whether there any clear segments of the disc margin or whether it is swollen all around. An isolated area of 'swelling' may actually be myelination of nerve fibres around the nerve head.
- Pallor suggests an additional range of conditions (compare to the other side). If swelling is severe, it may be hard to distinguish the disc from the background retina.
- Look for spontaneous venous pulsation: seen in the veins just as they emerge from the optic nerve. This is also absent in 20% of normal patients.
- Examine the rest of the fundus for other significant findings (eg, pallor, haemorrhages, abnormal-looking vasculature).
- Always examine both eyes.
Evaluation of optic nerve function

- Check visual acuity (VA) with a Snellen chart.
- Check for an RAPD using the swinging flashlight test:
  - Examine in a dark room, use a bright light source and ask the patient to gaze into the distance (such as a far wall) to avoid physiological constriction of the pupil and to maximise chances of spotting an abnormal response.
  - Shine the light source from one eye to the other in rapid succession.
  - Normally, both pupils should constrict briskly when the light is shone in either eye.
  - In the presence of an RAPD, stimulation of the normal eye elicits a brisk constriction of both pupils but when the light is shone on the diseased eye, both pupils dilate. The dilatation produced by withdrawing the light from the normal eye outweighs the weak constriction produced by shining light on the diseased eye - this is why it is called a relative afferent pupillary defect. (An afferent pupillary defect occurs where there is a critical optic nerve lesion or optic nerve transection: the patient is then blind in that eye - "it's all black when I cover my good eye" and neither pupil will constrict when the light is shone on the affected side).

- Check for colour impairment (dyschromatopsia) using Ishihara tests: cover the good eye first and move through the book, allowing about five seconds per number; then compare with the fellow eye. If the booklet is not available, ask the patient to compare the colour of a bright red object. Red desaturation is an early sign of optic nerve disease.
- Assess brightness sensitivity: shine a light in each eye and ask the patient to compare the brightness.
- Visual fields to confrontation, specifically looking for an enlarged blind spot.
- Assess the neurological system and the cranial nerves.

Investigation of optic disc swelling\(^[4]\)

- Urgent neuro-imaging; MRI with gadolinium enhancement is ideal.
- Further investigations may include magnetic resonance venography (MRV) to check the cerebral venous sinuses, lumbar puncture to check the opening pressure, CSF biochemistry and microbiology, and fluorescein angiography of the fundus.
- Ultrasonography or spectral domain coherence retinal tomography (provides data to form a 3D image) may play a role in diagnosing papilloedema, especially in differentiating from pseudopapilloedema.\(^{[5, 6]}\)

Causes of optic disc swelling\(^[7]\)

**Papilloedema**\(^{[2, 8]}\)

Papilloedema is optic disc swelling resulting from raised ICP: it is therefore almost always bilateral. The optic nerve sheath is continuous with the subarachnoid space, so that increased ICP is transmitted to the subarachnoid space surrounding the optic nerve. The anterior end of the optic nerve stops quite abruptly at the eye. The pressure prevents axonal flow back along the nerve, causing swelling and protrusion of the optic nerve at its head into the globe.

The time course for the development of papilloedema depends on the cause. It may be weeks if the rise in ICP is slow and mild, but it can occur within a day if the ICP rises suddenly and severely. If there is optic atrophy then there will be little or no papilloedema, even in the presence of raised ICP.

Patients with suspected papilloedema should be considered to have an intracranial mass until proved otherwise. Imaging of the brain and orbits is mandatory and a lumbar puncture may also be performed if imaging is normal. Not all patients with raised ICP develop papilloedema - this depends on the site and size of the tumour and, in infants with open fontanelles, it may fail to occur altogether. Patients who have previously had papilloedema may also fail to redevelop it in the future.
Presentation

- There may be few visual symptoms in early cases, although symptoms of raised ICP include headache (worse on waking, straining and bending), nausea and vomiting.
- Hypermetropia may increase due to changes in shape of the back of the eye. With chronicity, blurring of vision and eventually peripheral or complete visual field loss may be experienced.
- In contrast to other forms of disc swelling, VA is not impaired initially but in later stages there may be an increase in size of the blind spot.
- Severe papilloedema may cause transient episodes of visual loss, particularly when rising to stand (transient visual obscurations).
- There may be diplopia if there is a VI cranial nerve palsy.
- There may be a history of head trauma.
- Consider essential intracranial hypertension. 90% of cases occur in woman of child-bearing age, with raised BMI, taking the combined oral contraceptive pill.

Examination findings
The following are suggestive of papilloedema:

- **Ocular findings:**
  - Disc swelling - usually bilateral, sometimes asymmetrical.
  - Venous engorgement (typically the first sign in papilloedema).
  - Absent venous pulsation (may be absent in papilloedema).
  - Haemorrhages over or adjacent to the optic disc.
  - Blurring of optic margins.
  - Elevation of optic disc - if the disc is significantly swollen it may be hard to focus on the whole of it at the same time.
  - Radial retinal lines (Paton’s lines) radiating out from the disc.
  - Visual field defects - eg, an enlarged blind spot.
  - VA - may remain relatively intact in mild-to-moderate papilloedema and in many other causes of optic nerve swelling.
  - Impaired colour vision, red desaturation.
  - May have an RAPD or a VI cranial nerve palsy.

- **Systemic findings:**
  - Neurological signs depending on the cause of the raised ICP.

Classification of papilloedema\(^9,\,10\)
There are several proposed classifications of which the Frisen Scale (based on observation) is one of the most widely accepted classifications of severity. This scale grades papilloedema by the appearance of the optic disc borders, the diameter and degree of protrusion of the optic nerve head, the appearance of nearby blood vessels and the optic cup and signs of retinal and retinal nerve fibre disruption close to the disc:

- **Stage 0 - normal optic disc:** blurring of nasal, superior and inferior poles in inverse proportion to disc diameter. Radial nerve fibre layer (NFL) without NFL tortuosity. Rare obscuration of a major blood vessel, usually on the upper pole.
- **Stage 1 - very early papilloedema:** obscuration of the nasal border of the disc. No elevation of the disc borders. Disruption of the normal radial NFL arrangement with greyish opacity accentuating NFL bundles. Normal temporal disc margin. Subtle greyish halo with temporal gap (best seen with indirect ophthalmoscopy). Concentric or radial retrochoroidal folds.
- **Stage 2 - early papilloedema:** obscuration of all borders. Elevation of the nasal border. Complete peri-papillary halo.
- **Stage 3 - moderate papilloedema:** obscurations of all borders. Increased diameter of optic nerve head. Obscuration of one or more segments of major blood vessels leaving the disc. Peri-papillary halo has irregular outer fringe with finger-like extensions.
- **Stage 4 - marked papilloedema:** elevation of the entire nerve head. Obscuration of all borders. Peri-papillary halo. Total obscuration on the disc of a segment of a major blood vessel.
- **Stage 5 - severe papilloedema:** dome-shaped protrusion representing anterior expansion of the optic nerve head. Peri-papillary halo is narrow and smoothly demarcated. Total obscuration of a segment of a major blood vessel may be present. Obliteration of the optic cup.
Eye and optic nerve tumours
Optic nerve gliomas and optic sheath meningiomas are the principal tumours of the optic nerve. Optic nerve melanocytoma is rare. If the optic nerve is compressed or infiltrated then swelling may occur and vision may be affected.

See also separate *Eye and Optic Nerve Tumours* article.

- **Presentation:**
  - Reduced vision.
  - May complain of diplopia if globe movement is restricted.
  - Large lesions may also cause epiphora (tearing) and discomfort as a result of proptosis.
  - Sometimes, red eye due to congested blood vessels.
  - Gradual, painless fogging or dimming of vision (rarely, the tumour may bleed into itself causing sudden visual loss).
  - Meningiomas can also cause exophthalmos and an ipsilateral dilated pupil that does not react to direct light stimulation but might contract on consensual light stimulation.
  - Children may present with strabismus.
  - Optic nerve sheath meningiomas occasionally cause gaze-evoked amaurosis.
  - There can be overlap in symptoms with optic neuritis.

- **Ocular findings:**
  - VA reduced in later stages and there may be an RAPD.
  - Examination may reveal poor VA, loss of colour vision, visual field loss, optic disc swelling or optic atrophy.
  - Eye movement may be limited.
  - If the tumour is large, there may be proptosis ± limitation of eye movements.
  - Signs are usually unilateral unless there is chiasmal involvement.

- **Systemic findings:**
  - Depends on the nature of the lesion.

Optic neuritis
This refers to inflammation of the optic nerve, which can occur as a result of demyelinating or infective disease processes. The optic nerve head is occasionally swollen and usually pale. See also separate *Acute Optic Neuritis* article.

Demyelinating optic neuritis[11]

- **Presentation:**
  - Subacute monocular visual impairment (bilateral symptoms are unusual).
  - There may also be mild globe discomfort, particularly on moving the eye.
  - Demyelinating optic neuritis can occur in isolation but is more commonly part of a systemic problem which can include multiple sclerosis (MS), Devic’s disease (which is characterised by bilateral optic neuritis) and Schilder’s disease.

- **Ocular findings:**
  - Reduced VA.
  - RAPD.
  - Optic disc pallor.
  - Dyschromatopsia.
  - Visual field defects (most frequently altitudinal or arcuate).
  - Palsies involving the III, IV and VI cranial nerves may be present.
  - VA often returns over the course of several weeks (in 85% of cases, to 6/12 or better) but other functions often remain abnormal and a mild RAPD may persist.

- **Systemic findings:**
  - This depends on where demyelination has occurred.
Additional notes:
- There is a close association between optic neuritis and MS:
  - 16% of patients who have optic neuritis but a normal MRI scan will go on to develop MS within five years.
  - 50% of patients presenting with optic neuritis will have demyelinating lesions on MRI scan.
  - In 70% of established MS cases, there will be evidence of a previous episode of optic neuritis.
  - The risk of a patient who presents with optic neuritis developing subsequent MS increases with:
    - Winter onset.
    - HLA-DR2 positivity.
    - Worsening of symptoms with increase in body temperature (exercise, hot bath - Uhthoff's phenomenon).

Parainfectious optic neuritis
- Presentation:
  - Severe visual loss, usually bilateral, 1-3 weeks following a viral infection (eg, measles, mumps, chickenpox, whooping cough, glandular fever).
  - Children are more frequently affected than adults and this may occur after immunisations.
  - There may have been other neurological phenomena: headaches, ataxia or seizures.

Ocular findings:
- Disc may be swollen or normal.

Systemic findings:
- There may be features of meningo-encephalitis.

Additional notes:
- Spontaneous complete visual recovery is usual - a minority of patients will need systemic steroids if the visual loss is severe.

Infectious optic neuritis
Various infectious organisms can cause inflammation of the optic nerve head. These include:

Varicella-zoster virus:
- Primary optic neuritis is uncommon unless immunocompromised.
- Secondary optic neuritis arises from viral spread where acute retinal necrosis has occurred.
- Patients are treated with intravenous (IV) antivirals.
- See separate Chickenpox article.

Sinus infections:
- Occasionally, direct spread of infection (possibly due to sinus wall defects) or occlusive vasculitis may occur following spheno-ethmoidal sinusitis.
- Patients complain of severe headaches and recurrent episodes of unilateral visual loss.
- Treatment is with systemic antibiotics but surgical drainage of the sinus may also be needed.
- See separate Sinusitis article.

Cat-scratch fever:
- This self-limiting infection has a good prognosis with visual recovery occurring within 1-4 weeks of starting antimicrobial therapy.
- See separate Cat Scratch Disease article.
**Lyme disease:**
- Optic neuritis can occur.
- Peripheral neurological manifestations may mimic MS.
- A course of IV ceftriaxone is the management of choice.
- See separate Lyme disease article.

**Syphilis:**
- Acute optic neuritis may occur both in the primary or secondary stages.
- Involvement may be unilateral or bilateral.
- See separate Syphilis article.

**Ischaemic anterior optic neuropathy**[8]

**Non-arteritic**
Non-arteritic anterior ischaemic optic neuropathy is a partial or total infarction of the optic nerve head due to occlusion of the posterior ciliary arteries. Patients tend to be in the 45-65 age group and predisposing factors include hypertension, diabetes, hypercholesterolaemia, collagen vascular disease, antiphospholipid antibody syndrome, sudden hypotensive episodes and cataract surgery.

**Presentation:**
- Sudden painless monocular visual loss (often discovered on waking).

**Findings:**
- 70% of patients have moderate-to-severe reduction in VA
- Most will have a visual field defect (typically altitudinal).
- Dyschromatopsia proportional to the level of visual impairment.
- Pale oedematous disc.

**General management following referral:**
- Fasting lipids and glucose; exclusion of autoimmune diseases.
- Carotid artery Doppler ultrasound scans to rule out a source of emboli.
- Address cardiovascular risk factors.

**Prognosis:**
- Most patients experience no further reduction in VA but 30-50% go on to experience the same problem in the fellow eye within months or years.
- Non-arteritic anterior ischaemic optic neuropathy never occurs in the same eye twice.

**Arteritic (giant cell arteritis)**
See also separate Giant Cell Arteritis article.

Giant cell arteritis involves granulomatous inflammation of vessels involving the elastic tissues of the media and the adventitia. There is a predilection for the temporal, ophthalmic, posterior ciliary and vertebral arteries.

**Presentation:**
- Jaw claudication.
- Scalp tenderness.
- Neck pain.
- Malaise.
- Temporal artery tenderness.
- Visual reduction or loss.
- Age over 55 years of age.
- Episodes of amaurosis fugax may occur prior to infarction of the optic nerve head.
- Patients may also complain of flashing lights and periocular pain.
Findings:
- VA may be normal.
- Pale or swollen optic disc.
- Temporal artery may be tortuous and tender.

Initial management on suspicion of diagnosis:
- Giant cell arteritis is a medical emergency - failure to diagnose and treat adequately can result in severe sight impairment of one or both eyes.

Prognosis:
- Good if treatment is initiated promptly.
- Visual loss is irreversible but prompt treatment may save the other eye.

Toxic and nutritional optic neuropathies
These conditions may cause optic nerve swelling in the early stages. Toxic neuropathies are relatively uncommon in the developed world and are primarily associated with specific medications, occupational exposures, methanol, ethylene glycol, disulfiram, or tobacco and alcohol abuse. However, in developing nations, nutritional optic neuropathy is much more common, especially in regions afflicted by famine. Both genders and all races are equally affected and all ages are susceptible. The predominant cause of nutritional optic neuropathy is thought to be deficiency of B-complex vitamins. Those with pernicious anaemia are also at risk due to an impaired ability to absorb vitamin B12 from the intestinal tract.

Presentation:
- Visual loss is bilateral, symmetric, painless, gradual and progressive.
- Dyschromatopsia, particularly red desaturation, is a common early sign.
- Loss of VA may start with a blur or haze at the point of fixation, followed by a progressive decline.
- Vision loss can extend to total blindness but a loss beyond 20/400 is rare, except in the case of methanol ingestion.
- Peripheral vision is usually spared since the pattern of loss typically involves a central or cecocentral scotoma.

Findings:
- Pupils usually demonstrate a normal response to light and near stimulation.
- The optic disc may appear normal, swollen, or hyperemic in early stages. With hyperemia, disc haemorrhages may also be present.
- Continued damage to the optic nerve results in the development of optic atrophy, classically seen as temporal pallor of the optic disc.

Treatment:
- This is dictated by the cause of the disorder.

Prognosis:
- This is variable, depending on cause and degree.
- VA usually recovers before color vision.

Accelerated (malignant) hypertension
See separate Hypertensive Emergencies article.

Presentation:
- May be asymptomatic.
- Decreased VA.
- Episodes of temporary visual loss.
Ocular findings:
- Attenuation of arterioles (copper wiring).
- Arteriovenous nipping (narrowing of the veins as the arteries pass over them).
- Signs of vascular leakage (haemorrhages and exudates).
- Disc swelling occurs in the presence of very high blood pressure.

Systemic findings:
- Very high blood pressure (usually greater than 200 mm Hg systolic and/or 100 mm Hg diastolic).

Additional notes:
- Malignant hypertension is a medical emergency.
- It constitutes one of the most common causes of optic disc swelling.\(^{[12]}\)
- Although the mainstay of treatment is to lower the blood pressure, this must be done progressively, as a sudden drop can precipitate vascular occlusion.

Central retinal vein occlusion
See separate Retinal Vein Occlusions article.

Presentation:
- May be ischaemic or non-ischaemic.
- Occlusion of the central retinal vein results in a backlog and stagnation of blood, which leads to generalised (including disc) oedema.
- Painless reduction in VA which ranges from very mild to profound.
- This is monocular but the fellow eye can be affected.
- Rarely, patients complain of dull discomfort and a red eye.

Ocular findings:
- Mild-to-severe, sudden or progressive (over weeks) reduction in VA.
- If severe, RAPD may also be present.
- Dilated, tortuous veins.
- Diffuse retinal haemorrhages.
- Disc oedema.
- Later on, there may be conjunctival congestion.

Systemic findings:
- Evidence of cardiovascular disease.
- Other causes include clotting disorders, blood dyscrasias, paraproteinaemias, vasculitides, systemic lupus erythematosus and the oral contraceptive pill.

Additional notes:
- Patients tend to be managed conservatively with regard to the eye.
- There should be efforts made to address underlying predisposing factors.
- Some patients benefit from fluorescein angiography and laser treatment several months down the line.
- Prognosis for visual recovery is poor, particularly for the ischaemic form.

Diabetic papillopathy

Presentation:
- Progressive monocular visual loss.

Ocular findings:
- Modest decrease in VA (6/12 or better).
- Disc swelling may be unilateral or bilateral.
- Visual field defects (general constriction or scotoma), ± RAPD and dyschromatopsia.

Systemic findings:
- Evidence of peripheral diabetic vasculopathy.
Leber's optic neuropathy

**Presentation:**
- Unilateral, acute, severe, painless visual loss.
- Affects the second eye within two months.
- Typically affects males in their 20s (rarely, females aged 10-60 years).
- Patients may have family history, as this is an inherited disorder.

**Ocular findings:**
- Dilated capillaries on (swollen) disc surface.
- Visual field defects (scotoma).

**Systemic findings:**
- None.

**Additional notes:**
- Prognosis is generally poor, although a small number of patients will have some degree of recovery over months to years.

Pseudopapilloedema

Pseudopapilloedema is apparent disc swelling due to an underlying benign process. This could be due to colloid bodies (drusen) buried within the optic nerve head, an unusual angulation of the disc or a small disc from which the neurons emerge with a residual myelin sheath.

**Presentation:**
- Patients are usually asymptomatic with the finding incidental.
- Drusen can occasionally cause a progressive loss of peripheral vision (and, rarely, central vision).

**Ocular findings:**
- Apparent disc swelling (often bilateral) in the absence of any other findings.
- Small discs may look 'crowded' with the vessels sprouting out like a bunch of flowers.
- Residual myelin sheath is often limited to one part of the disc only.

The condition should be managed as true optic disc swelling until proven otherwise. The investigations outlined above should help guide the diagnosis. An ultrasound scan of the eye may identify the drusen; this can be confirmed using optical coherence tomography (OCT).[^13]
Management

If papilloedema is suspected (i.e., optic disc arising from raised ICP), there is an urgent need to rule out an intracranial mass.

Optic disc swelling that is not thought to be papilloedema should be referred according to the severity of the symptoms.

All patients with apparent optic nerve swelling should have an ophthalmological assessment and, unless an alternative diagnosis is clear, it is prudent to assume that the swollen disc is papilloedema until proven otherwise. Ultimately, the underlying cause needs to be addressed.

Further reading & references

12. Barnard S; Introduction to diseases of the optic nerve

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