

# Bilateral Optic Disc Swelling

**Disclaimer:** This Clinical Practice Guideline (CPG) was written for use in the Royal Victorian Eye and Ear Hospital Emergency Department. It should be used under the guidance of an ENT or Ophthalmology registrar. If clinical advice is required, please contact the Eye and Ear Admitting Officer for assistance: EYE: +61 3 9929 8033. ENT: +61 3 9929 8032. Links to internal Royal Victorian Eye and Ear Hospital documents cannot be accessed from the website CPG.

**SEE ALSO:** Idiopathic Intracranial Hypertension (IIH)

## Description:

Bilateral optic disc swelling refers to edema of the optic disc with concurrent increase in fluid within or surrounding the axons. The aetiologies of bilateral optic disc swelling include life-threatening (e.g. malignant hypertension) and/or reversible conditions (e.g. infectious neuropathy) which must be recognised and treated immediately. Papilloedema is a specific term referring to optic disc swelling due to proven raised intracranial pressure (e.g. idiopathic intracranial hypertension, brain tumour, etc.)

## How to Assess:

### Red Flags:

- Check blood pressure in all cases of optic disc swelling to rule out malignant hypertension (>180/120)
- Idiopathic intracranial hypertension (IIH) is a diagnosis of exclusion and can occur in patients with normal body weight. It should only ever be made in retrospect after the patient has been fully investigated (including MRI/MRV or CT/CTV and lumbar puncture (LP)) in all cases.
- In pre-existing optic atrophy, the disc may not be swollen despite raised intracranial pressure

## Definitions:

- **Papilloedema:** optic disc swelling due to proven raised intracranial pressure (ICP)
- **Pseudopapilloedema:** optic discs that appear elevated or have unclear margins without optic nerve axon swelling. See [Table 3](#) for the 2013 Revised Diagnostic Criteria for Pseudotumour Cerebri.
- **Pseudotumour cerebri:** refers to papilloedema with elevated intracranial pressure, in the absence of a structural abnormality (e.g. mass lesion) or CSF abnormality (e.g. meningitis)
  - **Idiopathic Intracranial Hypertension (IIH):** is sometimes used synonymously with pseudotumour cerebri, but it specifically applies to the case where there is no identifiable cause (e.g. no cerebral venous sinus thrombosis, use of vitamin A derivatives, etc.). Despite the use of the term "idiopathic", this particular syndrome is seen almost exclusively in overweight women of childbearing age, and should be used with caution in other demographics.
  - **Benign intracranial hypertension (BIH):** IIH, BIH term no longer used

## Aetiology

- Bilateral optic disc swelling can be caused by any of the following:
  - Papilloedema: space occupying lesion, hydrocephalus, meningitis, subarachnoid haemorrhage, dural venous sinus thrombosis, extracranial venous outflow obstruction, pseudotumour cerebri, IIH
  - Malignant hypertension
  - Infectious: syphilis, tuberculosis
  - Inflammatory: sarcoidosis, Vogt-Koyanagi-Harada syndrome
  - Nutritional/toxic optic neuropathy
    - Drugs: chloramphenicol, ethambutol, isoniazid, chloroquine
    - Toxins: methanol, lead, alcohol abuse
    - Vitamin deficiency: B12, B1 (thiamine), folate
  - Thyroid eye disease
- **Differential diagnoses**
  - Pseudopapilloedema
    - Optic disc drusen (ODD), high hypermetropic discs, vitreo-papillary traction, anomalous disc malformations (e.g. myelinated nerve fibres)
  - Arteritic or non-arteritic ischaemic optic neuropathy, sequential /bilateral

## On History

- Symptoms consistent with papilloedema and/or raised ICP:
  - Neurological
    - Headache (new or severe, worse in the morning or when coughing, straining, bending over or lying down)
    - Pulsatile tinnitus
    - Focal neurological symptoms related to causative lesion
  - Ophthalmic
    - Blurred vision, transient visual obscurations, photopsias, diplopia
- **Symptoms not consistent** with papilloedema:
  - Ocular pain, systemic symptoms e.g. cough, giant cell arteritis symptoms
- **Previous medical and surgical history consistent** with papilloedema and/or raised ICP:
  - Malignancy (possible metastases)
  - Deep vein thrombosis, pregnancy, miscarriages (possible coagulopathy causing dural venous sinus thrombosis)
  - IIH associated conditions: obesity, obstructive sleep apnoea, renal failure, anaemia, hypo/hyperthyroidism
- **Medications**
  - Medications associated with pseudotumour cerebri (e.g. vitamin A derivatives, tetracyclines, steroids, cyclosporin, oral contraceptive pills).

## On Examination

### General

- General inspection – obesity, overweight
- Vital signs – blood pressure (BP). Note malignant hypertension= BP>180/120. If hypertensive, must discuss with Emergency Registrar or St Vincent's Medical Registrar for investigation and management.

### Ophthalmic

- Assess for presence of optic neuropathy
- See [Table 2](#) for distinguishing characteristics of optic disc swelling vs pseudopapilloedema
- Visual acuity – may be normal
- Pupils – RAPD – may be subtle
- Visual fields – can be normal
- Colour vision (Ishihara plates test each eye separately)
- Complete anterior and posterior segment examination
  - Important to exclude ocular causes of disc swelling (e.g. uveitis, retinal vein occlusion, hypotony)
  - Prior to dilation, consider ongoing need to monitor pupil reactions. Only use tropicamide 0.5% or 1%
  - Disc examination
    - Colour, vasculature, margins, peripapillary retinal nerve fibre layer obscuring blood vessels
    - Grade papilloedema: see [Table 1](#), Modified Frisen Scale
    - Spontaneous venous pulsations (SVPs)
  - Present in 80% of normal subjects – generally indicate ICP less than 19cm CSF
  - Absence does not necessarily indicate raised ICP
    - Disc swelling in papilloedema is almost always bilateral, but may be asymmetrical
- Cranial nerve examination: extraocular movements – may be limited by orbital apex or pituitary tumour, may have limitation of abduction due to CN VI palsy from raised ICP
- Orbital exam: signs of carotid-cavernous fistula e.g. proptosis, conjunctival injection or chemosis, audible bruit

## On Investigation

- Papilloedema
  - MRI: urgent (within 24 hours) MRI optic nerves and brain with contrast
    - If MRI not available, same day CT Brain/Orbits and CT venogram
  - Lumbar puncture: to be obtained if no localizing neurological signs, and no evidence of a mass lesion on neuroimaging

- Organise through St Vincent's radiology. Request opening pressure, CSF biochemistry, microbiology and cytology, bloods for glucose and oligoclonal bands to compare with CSF
- Baseline automated visual field if presenting during business hours and optic nerve OCT if possible.
- If uveitis or a systemic cause of optic disc swelling is suspected:
  - Blood tests
    - Basic: FBC, UECs, LFTs, ESR, CRP
    - Inflammatory: ACE, ANA
    - Infectious: Syphilis serology, QuantiFERON Gold if risk of tuberculosis (where relevant)
  - Chest X-ray or CT Chest
  - Urinalysis (e.g. glycosuria in diabetes, haematuria in vasculitis)

## Acute Management

All cases of suspected papilloedema should be discussed with Neuro-ophthalmology consultant 'On-Call'

- Other relevant teams
  - St Vincent's Hospital
    - Neurology if suspect dural sinus thrombosis, meningitis
    - Neurosurgery if suspect brain tumour, hydrocephalus
  - Medical Retina/Ocular immunology if suspect retinal vein occlusion or uveitis
- Pseudotumour cerebri of known cause – manage underlying risk factor if possible and treat as per IIH
- IIH
  - Medical treatment:
    - Consider acetazolamide 250mg QID (contraindications are sulfur allergy, pregnancy, see below)
  - Surgical treatment is only a temporising measure to acutely save vision and/or relieve headaches in severe disease (high long-term failure rates):
    - Optic nerve sheath fenestration
  - Indication – vision-threatening disease where headache is not severe
  - Must discuss with neuro-ophthalmology and oculoplastics on-call
    - Neurosurgical shunt insertion
  - Indication – vision-threatening disease and/or severe headaches not controlled by medical treatment
  - Must discuss with neuro-ophthalmology and neurosurgery on-call at St Vincent's
  - Counsel patient regarding weight loss

- Special circumstances:
  - Pregnancy
    - No increased incidence of IIH during pregnancy and there is no increased risk of foetal loss
    - Acetazolamide should not be prescribed before 20 weeks gestation
    - Warn women not to become pregnant while taking acetazolamide and for 6 months after cessation
    - Rapid weight loss is contraindicated during pregnancy
  - Children/adolescents
    - A secondary cause of papilloedema is found in 50% of children/adolescents (e.g. space occupying lesion)

### Follow up:

- Discuss with neuro-ophthalmology consultant on-call regarding results, treatment and follow-up
- If patient is seen after-hours, they should be brought back to AOS within 24 to 48 hours for:
  - automated visual field testing
  - to ensure appropriate neuroimaging has been performed or is arranged
  - once neuroimaging has been reported, an LP must be organised through St Vincent's radiology where indicated.
- All cases must be discussed with neuro-ophthalmology on-call consultant
- Instruct patient to return to the Emergency Department if symptoms deteriorate.

*Table 1: Modified Frisen Scale*

Grade	Findings
0 – normal optic disc	
1 – minimal degree of oedema	C-shaped halo that is subtle and greyish with a temporal gap
2 – low degree of oedema	Circumferential halo
3 – moderate degree of oedema	Obscuration of $\geq 1$ segment of major blood vessels leaving the disc
4 – marked degree of oedema	Total obscuration on the disc of a segment of a major blood vessel on the disc
5 – severe degree of oedema	Obscuration of all vessels on the disc and leaving the disc

Table 2: Optic Disc Swelling vs Pseudopapilloedema: characteristics

Characteristic	Optic disc swelling	Pseudo-papilloedema
<b>Symptoms</b>	Present	Absent or present
<b>Visual acuity</b>	Generally affected	Preserved
<b>Hyperaemia</b>	Yes	No
<b>Disc abnormalities</b>	Capillary dilatation, telangiectasia, flame haemorrhages	May have visible drusen, myopic/tilted discs, hypermetropic discs
<b>Never fibre layer</b>	Oedematous, retinal vessels obscured	Normal, no obscuration of vessels
<b>Spontaneous venous pulsation</b>	Absent	May be present/absent
<b>OCT Disc (enhanced depth imaging - EDI)</b>	May have irregular hyper-reflective areas No hyper-reflective horizontal bands Retinal and/or choroidal folds	Optic disc drusen <ul style="list-style-type: none"> <li>○ always located above lamina cribrosa</li> <li>○ always have hypo-reflective core</li> <li>○ often have hyper-reflective margin</li> <li>○ may be associated with hyper-reflective horizontal bands</li> <li>○ No retinal or choroidal folds</li> </ul>
<b>B-scan</b>	No hyper-reflectivity	Hyper-reflectivity of calcified, superficial drusen
<b>Autofluorescence</b>	No autofluorescence	Autofluorescence (in superficial ODD)
<b>fundus fluorescein angiogram</b>	Disc leakage	No disc leakage, drusen may show hyperfluorescence
<b>Visual Fields</b>	Blind spot enlargement, generalised constriction, inferonasal loss	May also have blind spot enlargement with or without peripheral constriction and inferonasal loss
<b>Monitoring of changes with disc photos and OCT</b>	Progressive (days – months) change	Non-progressive

Table 3: 2013 Revised Diagnostic Criteria

**2013 Revised Diagnostic Criteria for Pseudotumour Cerebri**

- Papilloedema must be present
- Normal neurological examination except for cranial nerve abnormalities
- Neuroimaging: normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used
- Normal CSF composition
- Elevated LP opening pressure ( $\geq 25$  cmCSF in adults and  $\geq 28$  cmCSF in children [ $25$  cmCSF if the child is not sedated and not obese]) in a properly performed LP
- If papilloedema is not present, then there must either be a CN VI palsy (unilateral or bilateral) or  $\geq 3$  neuroimaging criteria satisfied:
  - Empty sella
  - Flattening of the posterior aspect of the globe
  - Distention of the perioptic subarachnoid space with or without a tortuous optic nerve
  - Transverse venous sinus stenosis

**References:**

1. Paine A, Burdon M, Miller N. The Neuro-Ophthalmology Survival Guide. Mosby Elsevier; 2006.
2. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology*. 2013 Sep 24;81(13):1159–65.
3. Jacks AS, Miller NR. Spontaneous retinal venous pulsation: aetiology and significance. *J Neurol Neurosurg Psychiatry*. 2003 Jan;74(1):7–9.
4. Wakerley BR, Tan MH, Ting EY. Idiopathic intracranial hypertension. *Cephalalgia Int J Headache*. 2015 Mar;35(3):248–61.
5. Malmqvist L, Bursztyn L, Costello F, Digre K, Fraser JA, Fraser C, et al. The Optic Disc Drusen Studies Consortium Recommendations for Diagnosis of Optic Disc Drusen Using Optical Coherence Tomography. *J Neuro-Ophthalmol Off J North Am Neuro-Ophthalmol Soc*. 2018 Sep;38(3):299–307.
6. Costello F, Malmqvist L, Hamann S. The Role of Optical Coherence Tomography in Differentiating Optic Disc Drusen from Optic Disc Edema. *Asia-Pac J Ophthalmol Phila Pa*. 2018 Aug;7(4):271–9.
7. Khandelwal N, Agarwal A, Kochhar R, Bapuraj JR, Singh P, Prabhakar S, et al. Comparison of CT venography with MR venography in cerebral sinovenous thrombosis. *AJR Am J Roentgenol*. 2006 Dec;187(6):1637–43.
8. Ehlers J, Chirag P. The Wills Eye Manual. Fifth. Lippincott Williams & Wilkins; 2008.
9. Scott CJ, Kardon RH, Lee AG, Frisén L, Wall M. Diagnosis and grading of papilledema in patients with raised intracranial pressure using optical coherence tomography vs clinical expert assessment using a clinical staging scale. *Arch Ophthalmol Chic Ill 1960*. 2010 Jun;128(6):705–11.

## Evidence Table

Author/s	Title	Source	Level of Evidence (I – VII)
Paine A, Burdon M, Miller NR	The Neuro-Ophthalmology Survival Guide	Mosby Elsevier, 2006	I
Friedman DI, Liu GT, Digre KB	Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children	Neurology. 2013;81(13):1159-65.	I
Jacks AS, Miller NR	Spontaneous retinal venous pulsation: aetiology and significance.	J Neurol Neurosurg Psychiatry. 2003;74(1):7-9.	III
Wakerley BR, Tan MH, Ting EY	Idiopathic intracranial hypertension.	Cephalalgia. 2015;35(3):248-61	I
Malmqvist L, Bursztyn L et al	The Optic Disc Drusen Studies Consortium Recommendations for Diagnosis of Optic Disc Drusen Using Optical Coherence Tomography	J Neuro-Ophthalmol Off J North Am Neuro-Ophthalmol Soc. 2018 Sep;38(3):299–307	VII
Costello F, Malmqvist L et al	The Role of Optical Coherence Tomography in Differentiating Optic Disc Drusen from Optic Disc Edema.	Asia-Pac J Ophthalmol Phila Pa. 2018 Aug;7(4):271–9	I
Khandelwal N, Agarwal A et al	Comparison of CT venography with MR venography in cerebral sinovenous thrombosis.	AJR Am J Roentgenol. 2006;187(6):1637-43.	III
Ehlers J, Chirag P	The Wills Eye Manual	Lippincott Williams & Wilkins; 2008.	I
Scott CJ, Kardon RH et al	Diagnosis and grading of papilledema in patients with raised intracranial pressure using optical coherence tomography vs clinical expert assessment using a clinical staging scale.	Arch Ophthalmol. 2010;128(6):705-11	III

## The Hierarchy of Evidence

The Hierarchy of evidence is based on summaries from the National Health and Medical Research Council (2009), the Oxford Centre for Evidence- based Medicine Levels of Evidence (2011) and Melynck and Fineout-Overholt (2011).

- I. Evidence obtained from a systematic review of all relevant randomised control trials.
- II. Evidence obtained from at least one well designed randomised control trial.
- III. Evidence obtained from well-designed controlled trials without randomisation.
- IV. Evidence obtained from well-designed cohort studies, case control studies, interrupted time series with a control group, historically controlled studies, interrupted time series without a control group or with case series.
- V. Evidence obtained from systematic reviews of descriptive and qualitative studies.
- VI. Evidence obtained from single descriptive and qualitative studies.
- VII. Expert opinion from clinician, authorities and/or reports of expert committees or based on physiology.



Version Details:	
CPG No:	CPG6.0
Responsible Executive:	Executive Director, Medical Services
Review Officer:	Clinical Practice Guideline Working Party
Contributors:	Head of Neuro-ophthalmology
National Standard(s):	Clinical Governance
Version Number:	1
Approval Date:	08/07/2020
Next Review Due:	07/07/2025