

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 Ophthalmology or ENT 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 Eye and Ear documents cannot be accessed from the website CPG.

See also: [Royal Children's Hospital Guidelines](#)

Description:

Bilateral optic disc swelling refers to oedema 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.)

Red Flags:

- Rule out malignant hypertension (BP >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.
- In pre-existing optic atrophy, the disc may not be swollen despite raised intracranial pressure (ICP)

How to Assess:

Definitions:

- **Papilloedema:** optic disc swelling due to proven raised ICP
- **Pseudopapilloedema:** optic discs that appear elevated or have unclear margins without optic nerve axon swelling.
- **Idiopathic Intracranial Hypertension (IIH):**
 - Papilloedema with elevated ICP, in the absence of a structural abnormality or abnormal cerebrospinal fluid (CSF). Elevated CSF opening pressure (OP) ($\geq 25\text{cmH}_2\text{O}$ in adults or $\geq 28\text{cmH}_2\text{O}$ in children) on lumbar puncture (LP). Note that if the opening pressure is $< 25\text{cmH}_2\text{O}$ in adults, but clinical symptoms and signs are consistent with IIH, then treat as possible IIH. An abnormal CSF should raise concern for a secondary cause of raised ICP.
 - 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. Atypical cases should raise suspicion for secondary causes.
 - Also known as pseudotumour cerebri and benign intracranial hypertension (BIH). BIH term no longer used

Aetiology

Bilateral optic disc swelling can be caused by any of the following:

1. **Primary:** idiopathic intracranial hypertension
2. **Secondary**
 - **Structural:** space occupying lesion, hydrocephalus, meningitis, subarachnoid haemorrhage, dural venous sinus thrombosis, extracranial venous outflow obstruction, spinal cord tumour
 - **Circulatory:** malignant hypertension, congestive heart failure
 - **Endocrine:** thyroid disease (Hypo- or hyper-), corticosteroid deficiency or excess, pituitary adenoma, hypoparathyroidism
 - **Infectious:** syphilis, tuberculosis, cryptococcus, bartonella, viral meningitis (EBV, Coxsackie B), lyme disease, HIV, brucella, dengue, West Nile virus, malaria, leptospirosis, coccidioidomycosis, parasitic disease
 - **Inflammatory:** sarcoidosis, Vogt-Koyanagi-Harada syndrome
 - **Neoplastic:** leukaemia, lymphoproliferative disorders, myeloma, POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, skin changes):
 - **Haematological:** anaemia (from any cause), polycythemia rubra vera, idiopathic thrombocytopenia purpura, paroxysmal nocturnal haemoglobinuria, monoclonal gammopathy
 - **Nutritional:** Vitamin B12, Vitamin B1, folate deficiencies, Vitamin A excess or deficiency
 - **Medications:** steroids, contraceptives, phenytoin, tetracyclines, nalidixic acid, amiodarone, Vitamin A derivatives, nitrofurantoin, sulfamethoxazole, lithium, chloramphenicol, ethambutol, isoniazid, chloroquine, cyclosporin
 - **Toxins:** methanol, lead, alcohol abuse

Differential diagnoses:

- Pseudopapilloedema
 - Optic disc drusen (ODD), high hypermetropic discs, vitreo-papillary traction, anomalous disc malformations (e.g. myelinated nerve fibres). See [Table 2](#).
- Arteritic or non-arteritic ischaemic optic neuropathy, sequential /bilateral

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 or other intracranial sounds
 - Any focal neurological symptom should raise suspicion of a secondary cause except a cranial nerve VI palsy or divergence insufficiency, which can be associated with raised ICP.
 - Meningeal irritation: nausea, vomiting, photophobia
 - Ocular: blurred vision, transient visual obscurations, photopsias, diplopia. Be suspicious of other causes if there is early central visual loss.
- Past medical and surgical history consistent with papilloedema and/or raised ICP:
 - Malignancy (i.e. possible metastases), deep vein thrombosis, pregnancy, miscarriages (possible coagulopathy causing dural venous sinus thrombosis)
 - IIH associations: women of childbearing age, obesity or recent weight gain, obstructive sleep apnoea, renal failure, anaemia, hypo/hyperthyroidism
- Medications (as above)

Examination:

General:

- Overweight/obesity
- Check blood pressure: malignant hypertension= BP>180/120. Discuss with Emergency Registrar or St Vincent's Medical Registrar for investigations and management.

Ocular:

See [Table 2](#) for distinguishing optic disc swelling vs pseudopapilloedema

- Visual acuity: may be normal. Loss of vision is major morbidity in IIH and can be gradual or acute
- Pupils: Relative afferent pupillary defect (RAPD) (may be subtle)
- Visual fields: may be normal. Increase blind spot common.
- Colour vision
- Complete anterior and posterior segment examination
 - Exclude ocular causes of disc swelling (e.g. uveitis, retinal vein occlusion, hypotony)
 - Disc examination
 - Colour, vasculature, margins, peripapillary retinal nerve fibre layer obscuring blood vessels
 - Grade papilloedema: see [Table 1](#), Modified Frisen Scale

- Disc swelling in papilloedema is almost always bilateral, but may be asymmetrical or unilateral
- Spontaneous venous pulsations (SVPs): present in 80% of normal people – generally indicate ICP less than 19cmH₂O, but absence does not always mean raised ICP
- Cranial nerve (CN) examination: extraocular movements may be limited by orbital apex or pituitary tumour; may have CN VI palsy (unilateral or bilateral) from raised ICP
- Orbit exam: exclude carotid-cavernous fistula e.g. proptosis, conjunctival injection or chemosis, audible bruit

Investigations

- Baseline automated Humphrey visual field and optic nerve OCT. Document confrontational visual fields if after hours.
- MRI and MRV brain/orbits with contrast: urgent (within 24 hours)
 - If MRI not available, same day CT Brain/Orbits and CT venogram
 - Children
 - St Vincent's Private Radiology will not use contrast in children < 14 years old and may not be able to image children who are unable to remain still
 - Consider referral to Royal Childrens Hospital (RCH) if not suitable for imaging at St Vincent's Private Radiology
 - MRI Findings
 - Papilloedema: optic nerve sheath distension, posterior flattening of the globe
 - IIH: empty sella turcica, unilateral or bilateral transverse sinus stenosis
 - Other aetiology: intracranial mass, subdural collection, meningeal disease
 - Lumbar puncture: if no localising neurological signs, and normal 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
 - For children <16 years old with optic disc swelling with red flags: disc photos and retinal nerve fibre OCT

Acute Management:

- All cases of suspected papilloedema should be discussed with on-call Neuro-ophthalmology consultant
- Contact
 - Medical Retina/Ocular Immunology Fellow if associated uveitis
 - St Vincent's Hospital neurology registrar on-call if dural sinus thrombosis (confirmed or suspected), meningitis or systemic cause suspected
 - St Vincent's Hospital neurosurgery on-call if confirmed brain tumour, hydrocephalus
- IIH
 - Discuss all newly diagnosed IIH patients with Neuro-ophthalmology consultant on-call
 - Non-medical treatment: counsel patient regarding weight loss
 - Medical treatment:
 - Consider acetazolamide 500mg BD (contraindications are sulfa 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) with two options:
 - Optic nerve sheath fenestration
 - Indication: vision-threatening disease where headaches 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
- Special circumstances:
 - Pregnancy
 - No increased incidence of IIH during pregnancy
 - Acetazolamide should not be prescribed < 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
 - For children <16 years old:
 - Incidental finding of optic disc elevation or swelling and no red flags for raised ICP, discuss with RCH ophthalmology registrar on-call for an outpatient clinic (or refer privately for follow-up)
 - Optic disc swelling with red flags: disc photos and disc RNFL OCTs. Discuss with RCH on-call neurology and ophthalmology, then refer to RCH emergency department. Forward photos/ OCT images to RCH

Follow up:

- Discuss with Neuro-ophthalmology consultant on-call regarding results, treatment and follow-up (follow-up for associated uveitis, systemic disease, severe headaches or vision loss in IIH and children as per above)
 - If patient is seen after-hours, they should be brought back to AOS within 24 to 48 hours for Humphrey visual field, and to ensure appropriate neuroimaging has been performed. If neuroimaging is normal, organise a LP with St Vincent's Private Radiology.
- 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 ≥ 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
Symptoms	Present	Absent or present
Visual acuity	Generally affected	Preserved
Hyperaemia	Yes	No
Disc abnormalities	Capillary dilatation, telangiectasia, flame haemorrhages	May have visible drusen, myopic/tilted discs, hypermetropic discs
Never fibre layer	Oedematous, retinal vessels obscured	Normal, no obscuration of vessels
Spontaneous venous pulsation	Absent	May be present/absent
OCT Disc (enhanced depth imaging - EDI)	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"> ○ always located above lamina cribrosa ○ always have hypo-reflective core ○ often have hyper-reflective margin ○ may be associated with hyper-reflective horizontal bands ○ No retinal or choroidal folds
B-scan	No hyper-reflectivity	Hyper-reflectivity of calcified, superficial drusen
Autofluorescence	No autofluorescence	Autofluorescence (in superficial ODD)
fundus fluorescein angiogram	Disc leakage	No disc leakage, drusen may show hyperfluorescence
Visual Fields	Blind spot enlargement, generalised constriction, inferonasal loss	May also have blind spot enlargement with or without peripheral constriction and inferonasal loss
Monitoring of changes with disc photos and OCT	Progressive (days – months) change	Non-progressive

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.

Appendix 1

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.
10. Kidd DP, Newman N and Biousse V. Neuro-ophthalmology. Butterworth-Heinemann Elsevier, Philadelphia, 2008

Version Details:	
CPG No:	CPG6.0
Responsible Executive:	Executive Director, Medical Services
Review Officer:	Director, Emergency Department
Contributor(s):	<ul style="list-style-type: none"> • Clinical Practice Guideline Working Group • Head of Neuro-Ophthalmology Unit
National Standard(s):	Comprehensive Care
Version Number:	2.0
Approval Date:	07/06/2021
Next Review Due:	07/06/2026