CLINICAL PRACTICE GUIDELINE: Emergency Department

Optic Neuritis

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: Acute Loss of Vision, Visual Field Loss, Multiple Sclerosis

Description:

Optic Neuritis (ON) is an acute inflammation of the optic nerve. It is traditionally divided into 'typical' and 'atypical' presentations. 'Typical' optic neuritis generally has a good prognosis for recovery of vision, and is usually a demyelinating event. It can occur in isolation ('idiopathic'), or as part of the presentation or relapse of a demyelinating disorder like multiple sclerosis (MS). 'Atypical' optic neuritis is characterised by the presence of 'red flag' variations in presentation and raises the possibility of a different cause (e.g. infections, autoimmune). Recognition of these atypical features necessitates a more detailed clinical search for an underlying cause.

Red Flags:

- Typical optic neuritis is the only optic nerve inflammation that can be diagnosed and treated by history and examination alone, but the diagnosis can only be confirmed retrospectively, once the patient's vision recovers over time. Typical optic neuritis carries an increased risk of MS and may be the first presentation of MS.
- Beware of 'atypical optic neuritis/neuropathies' that warrant further investigations. See Table 1 for features of atypical optic neuritis. Atypical optic neuritis should be recognised as early as possible as some of these patients are at increased risk of severe visual loss

How to Assess:

Typical Optic Neuritis:

- Age typically 15-50 years of age (male or female)
- Unilateral, acute (within days to two weeks) visual loss. Improvement in visual acuity (VA) after four weeks from onset of symptoms
- Reduced colour sensitivity
- Pain in or behind the eye that worsens with eye movements in most patients
- No diplopia
- Other symptoms: Pulfrich phenomenon - altered depth perception of moving objects, and Uhthoff phenomenon – temporary worsening of visual symptoms when body temperature increases or with exercise.
- No other neurological complaints at time of presentation in most cases, although at times patient may report previous episodes of neurological dysfunction (e.g. limb weakness or numbness) that had resolved.
- Otherwise systemically well: no history of cancer, vasculitis or autoimmune disease

Table 1: Features of typical and atypical presentations of optic neuritis

	Typical	Atypical
Age	Young adult patient <50 years old	Age >50 or <12 years
Race	Caucasian	Non-caucasian
Optic neuritis	Unilateral	Bilateral or recurrent
Visual loss	Acute or subacute visual loss Progressive over a few days up to 2 weeks Unilateral visual loss with reduced colour and contrast vision, any type of visual field defect	Sudden visual loss Progressive visual loss for >2 weeks Severe visual loss to no perception of light Bilateral visual loss
Pain	Periocular pain and painful eye movements	No pain Severe or persistent pain >2 weeks
Systemic symptoms	Neurological signs and symptoms suggestive of MS	Clinical symptoms suggestive of diseases other than MS (neuromyelitis optica (NMO), connective tissue disorders, sarcoidosis, vasculitis)
Medical History	Previous history of ON or MS	Previous history of neoplasia
Optic nerve	Normal or swollen optic disc	Optic atrophy without history of ON or MS Optic disc haemorrhage Severe optic disc oedema
Retinal abnormalities	Normal macula and peripheral retina	Macular star
Other ocular findings	Uveitis or retinal periphlebitis possible	Marked uveitis or retinal periphlebitis
Steroids	No deterioration after withdrawal of steroids	Deterioration after withdrawal of steroids
Recovery	Spontaneous improvement after 2–3 weeks	Absence of recovery >3 weeks after onset

Examination:

Document the following parameters on examination in both eyes: VA, RAPD, light saturation, confrontational visual field (VF), colour vision (Ishihara colour plates and red colour saturation), pursuit, saccades and vergence including cover/uncover and alternating cover tests.

- Reduced VA (6/6 to 'no perception of light')
- Reduced colour vision (Ishihara and red colour saturation)
- RAPD and reduced light saturation
- Any VF defect: most commonly a diffuse loss including central/centro-cecal scotomas, but any type of field loss can be associated with optic neuritis.
- Optic disc appearance: approximately two thirds of patients have a normal-appearing disc (retro-bulbar optic neuritis); one third can have mild to moderate disc swelling with no cotton wool spots or hard exudates. Large peri-papillary haemorrhages are less likely.
- Intraocular inflammation: a minority of cases with typical optic neuritis can present with signs of pars planitis. This presentation warrants further investigation to rule out causes, e.g.: sarcoidosis, cat scratch disease, syphilis and vasculitis.
- No proptosis, no ptosis, normal eye movements, normal corneal and facial sensation

Investigations:

- Humphrey VF: any type of field loss/defect (as above) in affected eye. Unaffected eye may demonstrate asymptomatic field loss (up to 75% of cases).
- MRI brain and orbits: often shows features consistent with the diagnosis (increased T2 signal within the affected optic nerve, and sometimes contrast enhancement). The principal role for MRI is determining the risk of MS: presence of additional demyelinating lesions is the strongest predictive tool. It is standard practice to request a (non-urgent) MRI brain and orbits. In atypical presentations MRI should be requested for the brain and orbits with contrast - the urgency of this depends upon the clinical tempo of the presentation.
- Fundus photographs and OCT: may be helpful in certain cases, particularly if there is disc swelling or suspicion of macular disease. Retinal nerve fiber layer (RNFL) thickness and Ganglion Cell Layer analysis can be useful in evaluating clinical or subclinical optic atrophy.
- For atypical presentation of optic neuritis consider further investigations: laboratory testing, Chest X-ray/CT (for sarcoidosis and tuberculosis), lumbar puncture. (See Table 2)
- No evidence to suggest a need for routine blood tests or CSF examination in patients with typical optic neuritis.

Table 2: Laboratory testing

Full blood examination (FBC) for atypical optic neuritis
ESR, CRP
Syphilis serology
Serum angiotensin-converting enzyme (ACE), calcium
Anti-nuclear antibodies (ANA)
NMO-IgG and MOG-Ab
ANCA
Quantiferon gold testing
Anti-Bartonella henselae antibodies (Cat scratch)
LP (if necessary) to check opening pressure and CSF (for microscopy, culture and sensitivities), protein, glucose, cytology, flow cytometry, and oligoclonal bands

Acute Management:

- There is no evidence of long-term benefit with either oral or intravenous corticosteroids compared to placebo in regards to VA, contrast sensitivity or VF loss according to Cochrane Database. According to the Optic Neuritis Treatment Trial (ONTT) 93% of patients will have recovery of vision to at least 6/12.
- If a patient presents with typical optic neuritis within 14 days of onset of symptoms, a course of high dose IV methylprednisolone should be considered if there is: severe visual loss (<6/12), white matter lesions on MRI or an occupational need for rapid return of vision.
- Intravenous 1 gram of methylprednisolone for three consecutive days (see Methylprednisolone Inpatient Procedure). An oral prednisolone taper is optional. Atypical optic neuritis eg. Neuromyelitis optica spectrum disorder (NMOSD), may be refractory to standard steroid treatment and may require escalation including plasma exchange. Conversely, MOG and sarcoid may require a prolonged oral taper. These cases should be discussed with neuro-ophthalmology to guide management.
- Oral prednisolone alone was found to be harmful and should not be offered as initial treatment. It did not speed up recovery and actually increased the likelihood of recurrent optic neuritis (ONTT).

Follow up:

- New presentations with presumed acute optic neuritis should be discussed with the on-call Neuro-ophthalmology consultant to determine appropriate management, investigations and follow-up arrangements.
- VF should be requested at baseline to enable comparison at follow-up visits.
- Patients should have a review appointment arranged (Neuro-ophthalmology Clinic) after 4 weeks to check for early signs of recovery as anticipated. Patients with atypical features, and/or severe vision loss, may require more urgent review.
- Patients should be instructed to return to the Emergency Department if vision deteriorates.

If evidence of progression or atypical features, further investigations are required as previously outlined (see Table 2) and in consultation with the Neuro-ophthalmology team.

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
	The Wills Eye Manual, 5th edition		I
Gal RL, Vedula SS, Beck R	Corticosteroids for treating optic neuritis.	Cochrane Database Syst Rev. 2012 Apr 18; 4: CD001430.	I
Optic neuritis Treatment Trial (ONTT) 1992-2006 Beck RW, Cleary PA, Andersen MMJ, Keltner JL, Shults WT, Kaufman DI, et al.	A randomized controlled trial of corticosteroids in the treatment of acute optic neuritis. The Optic Neuritis Study Group.	New England Journal Of Medicine 1992; 326(9):581–8.	II
Axel Petzold, Mike P. Wattjes, Fiona Costello, Jose Flores-Rivera, Clare L. Fraser, Kazuo Fujihara, Jacqueline Leavitt, Romain Marginier, Friedemann Paul, Sven Schippling, Christian Sindic, Pablo Villoslada, Brian Weinshenker and Gordon T. Plant.	The investigation of acute optic neuritis: a review and proposed protocol.	Nature Reviews Neurology 2014; 10: 447–458	I
Dumitrascu O and Gordon LK	Atypical Optic Neuritis	Focal Points AAO 2014; Vol XXXII (9)	
Voss E, Raab P, Trebst C, Stangel M	Clinical approach to optic neuritis: pitfalls, red flags and differential diagnosis	Ther Adv Neurol Disord. 2011 Mar; 4(2): 123–134	VII

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 Melynk 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.

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