

Ocular electrophysiology

A 34-year-old woman, who is a CEO in a multinational firm, has been losing vision over the last 12 months. She has seen her opticians, who initially tried different glasses but could not improve things. Clinical examination is unremarkable. How would you investigate / manage this patient?

The clinical presentation is suggestive of progressive visual loss due to non-refractive causes in a young woman. Further clarification needs to be sought regarding the nature and progression of visual loss, as well as presence of associated ocular and systemic symptoms. Detailed medical history should be taken to rule out possible systemic causes for the visual loss. Assessment of dietary intake, smoking and alcoholism could provide pointers towards the underlying cause. Implication of visual loss related to work and driving must be assessed.

History pertaining to visual loss

- Was the vision loss unilateral / bilateral?
- If bilateral, was the visual loss simultaneous or consecutive?
- Is the distance / near vision / both affected?
- Is the vision affected during the day / night or both?
- Was it an acute onset event with progressive / stepwise decrease or was it gradual decrease from the onset?
- Is the visual field intact?
- Is the field loss unilateral or bilateral?
- Is the visual defect central / peripheral / generalised?
- Any abnormalities with colour vision / contrast?
- Is metamorphopsia / micropsia / macropsia present?

- Other associated symptoms like flashes / floaters?
- Does the patient complain of glare / dry eye symptoms?
- Any neurological symptoms?
- Is the visual loss associated with any other systemic symptoms?

Medical history

- Systemic diagnosis?
- Specifically enquire about history of malignancy and treatment with radiotherapy / chemotherapy.
- Has the patient been treated with any medications that could cause optic neuropathy / maculopathy?

Family history: Detailed family history – macula / retinal dystrophies, hereditary optic neuropathy.

Dietary history: Assessment for poor nutritional intake.

Habits: History to assess severity and chronicity for smoking and alcoholism.

Occupation

- What are the visual requirements for the job?
- How is the patient currently coping with the visual needs? Visual aids?

Driving: Still driving or not?

Examination

1. Colour vision
 2. Visual fields – confrontation fields, Amsler
 3. Pupils – relative afferent pupil defect (RAPD)
 4. Ocular motility
 5. Anterior segment examination – ocular surface problems / media opacity
 6. Intraocular pressure
 7. Vitreous? Vitritis
 8. Fundoscopy
Disc – oedema / pallor / cupping / neuro-retinal rim / shunt vessels
Macular / retinal pathology
 9. Measure blood pressure.
- A methodical history and clinical

examination approach will usually give sufficient information to anatomically localise the cause for visual loss on the basis of which subsequent investigations should be tailored.

Investigations

Neuro-ophthalmic causes:

- Goldmann visual perimetry
- Magnetic resonance imaging (MRI) brain and orbit with contrast, magnetic resonance venography
- Bloods – full blood count, erythrocyte sedimentation rate, C-reactive protein, liver function test, urea and electrolytes, angiotensin-converting enzyme, vasculitic screen, venereal disease research laboratory, B12 and folate levels
- Chest x-ray
- Lumbar puncture
- OCT
- Electro diagnostics (EDT).

Macular / retinal:

- OCT
- Fundus fluorescein angiography
- EDT.

Whilst definitive management will depend on the underlying diagnosis, care needs to be taken to ensure that visual rehabilitation measures are provided. The patient should also be informed as to whether they meet the DVLA requirements or not.

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TAKE HOME MESSAGE

Unexplained loss of vision requires a good history, electrophysiological testing and neuroradiological imaging to make an accurate diagnosis.

Ocular electrophysiology is an objective means of assessing retinal, macular, optic nerve and cortical visual function, each of which could explain this patient's symptoms.

Tests consist of the visual evoked potential (VEP), flash electroretinogram (ERG), pattern ERG (PERG), electrooculogram (EOG) and multifocal ERG or VEP (MfERG, MfVEP).

All electrophysiological investigations should be performed according to International Standardised Methodology (ISCEV) Standards in order that each centre performing ocular electrophysiology carries out similar stimulus and recording parameters compared to every other centre.

VEP: Flash stimulation assesses the functional integrity of the visual association areas of the visual cortex. Pattern stimulation is performed using a reversing checkerboard stimulus of different check sizes. It stimulates ganglion receptor fields within the central retina (magnocellular and parvocellular) and will give an assessment of the functional integrity of the central visual pathways from the central retina, along the optic nerve and the rest of the anterior visual pathways including the primary visual cortex.

VEPs can be useful in detecting optic neuropathy, but in reality any pathology affecting the central visual pathways can affect the amplitude and latency (implicit time) of the VEP. Delays of the pattern VEP beyond 135msec with responses of near normal amplitude would suggest optic neuropathy and / or demyelination of the optic nerve. Small check pattern responses can be useful in determining visual acuity.

By using multiple electrodes and stimulation of hemifields post chiasmal function or hemispheric abnormalities can be detected, including hemianopic field loss or albinism.

Flash ERGs: The Flash ERG should be recorded under scotopic and photopic conditions using a Ganzfeld stimulator. Dim flash scotopic stimulation will give a rod isolated response, whereas 30Hz photopic stimulation will give a cone isolated response.

The ERG consists of an a and b wave, the a wave from the photoreceptors, and the b wave from the bipolar and muller cells.

EOG: A light rise of >160% should be seen in normal individuals. The EOG is abnormal in inherited rod cone dystrophy, and toxic retinopathy, as well as some inherited maculopathies, juvenile Bests

Causes for gradually progressive visual loss:

1. **Optic neuropathy / chiasmal**
 - a. Compressive-meningioma, pituitary adenoma
 - b. Papilloedema
 - c. Inflammatory
 - Non-infectious – sarcoidosis, vasculitis
 - Infectious – tuberculosis, syphilis
 - d. Toxic – tobacco, ethambutol, amiodarone, bush tea, cassava
 - e. Nutritional – B12, folate, thiamine, riboflavin
 - f. Autoimmune optic neuropathy
 - g. Para-neoplastic optic neuropathy
 - h. Carcinoma / melanoma associated retinopathy
2. **Macular**
 - Macular dystrophies / other maculopathies
3. **Retina**
 - Retinal dystrophies
4. **Uveitis**
 - Intermediate / posterior uveitis
5. **Functional visual loss.**

disease, pattern dystrophy, North Carolina maculopathy.

Pattern ERG: Recorded under photopic conditions using a reversing checkerboard stimulus the PERG consistent of a P50 (preganglionic) and an N95 (ganglionic) response. The test is particularly useful in detecting maculopathy (P50) or optic atrophy (N95).

MfERG/VEP: Assesses macular and ganglion cell function over an array of visual field locations. It is useful in determining whether a field defect is due to retinal or optic nerve disease, and in determining progression.

Main uses of the ERG

Rod cone dystrophy: The ERG becomes subnormal at an early stage of the disease, particularly the rod isolated response.

Cone rod dystrophy: The cone 30Hz responses are absent whereas the scotopic responses are present although markedly reduced.

Cone dystrophy: The photopic and 30Hz responses are reduced or absent with preservation of scotopic responses.

Inflammatory or ischaemic retinopathy: The a waves are preserved but the b waves are reduced.

Birdshot chorioretinopathy: Delay of 30Hz b wave peak time indicates disease progression, whereas shortening of the latency is useful in determining the effectiveness of treatment.

Occult retinal disease: Unilateral delay of b wave peak time in the presence of a normal fundus appearance.

MEWDS: Negative or delayed b waves occur in the presence of flecks.

Stargardt's disease: ERGs are generally normal in this condition unless advanced, but PERGs are always abnormal.

Vitamin A retinopathy: Loss of the rod isolated response and reduction of the a waves is indicative of this condition.

Congenital stationary night blindness: Negative b waves is typical of the condition in its x-linked or recessive inheritance. If only the scotopic ERG is affected this indicates complete congenital stationary night blindness (CSNB), if both the scotopic and photopic ERG are affected then this is the incomplete form.

Fundus albipunctata: Scotopic ERG is grossly reduced but improves to almost normal values after 12 hours dark adaptation.

Dominant CSNB: (Nougaret) scotopic and photopic ERG are of identical amplitude and peak time.

Problems with interpreting pattern VEP

The pattern VEP is affected by poor fixation, incorrect refraction, pre-existing amblyopia and opaque media, and caution should be taken if any of these exist.

Macular disease can affect the latency of the VEP as much as optic neuropathy so caution would be recommended when suggesting that a delayed VEP indicates an optic neuropathy. A macular ocular coherence tomography (OCT) and PERG should always be performed in addition to VEP if optic neuropathy is suspected.

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