Visual disturbances

Gordon T Plant

Abstract

The eyes, ocular motor system and visual pathways of the central nervous system are frequent sites of neurological disease. As patients with visual symptoms often present first to an ophthalmologist or optometrist, it is as important for these two specialists to have some knowledge of neurological disorders affecting vision as it is for the neurologist and general physician. This article outlines the basic assessment of a patient presenting with impaired vision, either as a primary complaint or as part of a more generalized neurological or systemic disorder.

Keywords Cortical blindness; diplopia; hemianopia; optic neuropathy; pupil; visual aura; visual disturbance; visual loss

Key points

- When assessing a patient with visual loss, the history is of paramount importance
- Sudden onset points to a vascular cause, gradual onset to a compressive lesion
- In cases of transient visual loss, the duration of the episodes often points to the diagnosis
- Testing of visual acuity, colour vision, visual fields and pupillary light reflex usually provides evidence of the anatomical location of the lesion in the visual pathway
- The relative afferent pupil defect detects unilateral or asymmetrical optic neuropathy, which is therefore of no value in diagnosing bilateral symmetrical optic neuropathy
- The diagnosis is usually confirmed by neuro-imaging of the visual pathway
- Retinal imaging (optical coherence tomography) and visual electrodiagnostic testing are most useful in identifying retinal pathology that may be mistaken for optic neuropathy
- In assessing eye movement disorders and diplopia, it is important to master the following techniques in addition to examining the range of movements: the cover test, saccadic eye movements, smooth pursuit eye movements and the vestibulo-ocular reflex.

Visual loss

When assessing a patient complaining of deteriorating vision, the first task is to decide whether the impairment is monocular or binocular, whether the onset was gradual or abrupt and, with an abrupt onset, whether the impairment was transient or persistent. This is not always straightforward. A patient may not notice slowly progressive monocular visual loss until they happen to cover the unaffected eye, giving a pseudo-abrupt onset. Patients with transient homonymous hemianopia often report that the loss affected only the eye ipsilateral to the field defect. Patients with gradually progressive or congenital homonymous hemianopia may be unaware of the visual field loss until a perimetric examination is performed for another reason.

Visual loss is most likely to be reported if abrupt in onset, if it affects central vision and, especially, if it affects both eyes. Extensive bilateral visual field loss with no corresponding regions of loss in the two eyes (such as pure bitemporal hemianopia) can go unnoticed.

Examination in cases of visual failure

The first task is to decide whether one or both eyes are affected and whether the problem is caused by impaired optical quality or a disorder of the retina, the optic nerve, the chiasm or the post-chiasmal visual pathways. Inspection of the eye may reveal a paralytic strabismus or mechanical displacement of the globe itself (e.g. proptosis). Examination continues with an assessment of visual acuity, taking suitable precautions to ensure that neither a refractive error nor a lens opacity is a confounding factor.

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Visual acuity is impaired in most cases of optic neuropathy but spared in pure bitemporal and unilateral homonymous hemianopia. This is because visual acuity testing examines only central (foveal) vision, whereas extensive peripheral field loss — including hemianopia — spares acuity, although in hemianopia the patient may miss letters on one side of the chart.

Distance and near (reading) vision should be tested routinely with and without the patient's own glasses and with a pinhole (which compensates, at least in part, for any additional refractive error).

Patients with right homonymous hemianopia have specific difficulty with reading fluency (not acuity) because they cannot scan ahead. This occurs only when the macula is split and is referred to as 'hemianopic alexia'.[1] Patients with posterior left hemisphere lesions may also have 'acquired alexia' and may no longer be able to read word shapes but will read slowly and laboriously letter by letter.

Colour vision

Colour vision testing is of particular importance in neurological visual disorders as it is disproportionately affected in optic neuropathy.

The Ishihara pseudo-isochromatic plates are used as standard. There is some support for the use of smartphone apps of the

Ishihara plates. It is important to note that these plates distinguish the various degrees of Daltonism (red–green colour-blindness), although they test only for protanopia ('red' deficiency) and deuteranopia ('green' deficiency); there is no plate to test for tritanopia ('blue' deficiency). Therefore the detail of the errors made is not as significant in optic neuropathy, where various patterns of impaired colour vision can be found. However, it is clearly important to be able to recognize that a patient has Daltonism (the errors made are identical in each eye) when interpreting the findings.

It is usual to record whether or not the patient reads the first (control) plate as no colour vision is required for this. If the patient fails, there is no value in proceeding – it usually means that the patient's visual acuity is not adequate to read the plates (it is 6/60 or worse) but there are other explanations. For example, the disorder may be functional or patients with cortical visual disorders may not be able to perform the 'figure–ground' nature of the task (i.e. the ability to group the dots to perceive the number). Such patients will fail all the plates – including the control. This is often an early clinical finding in posterior cortical atrophy (a form of Alzheimer's disease that affects the visual cortex) and may be misinterpreted as loss of colour vision.

Colour vision can be assessed at the bedside by asking the patient to report any change in colour appearance of a coloured target. In the simplest form of this test, the patient is asked to compare the appearance of a small coloured target (e.g. a red pin head) foveally, viewing first with one eye and then the other. Patients with unilateral optic neuropathy usually report that the target looks paler (less saturated or closer to white) with the affected eye, although sometimes it may appear darker (maroon or brown). If both eyes are affected, this interocular comparison is not possible.

Contrast sensitivity is increasingly employed in the assessment of visual function in optic nerve disease. Charts on which the test letters change in contrast rather than size are used for this.

Visual fields

Visual field testing is a valuable skill that should be part of everyday practice for ophthalmologists, neurologists and other physicians. Begin with both the patient's eyes open and test in all four quadrants for the ability to see a moving finger and count fingers. This will exclude a gross homonymous hemianopia, and it is possible to look for visual inattention by testing simultaneously in the two hemifields. Then test monocularly in the same way in the four quadrants. If there is no abnormality, a small coloured target (e.g. a 4-mm red pin head) can be used to look for evidence of colour desaturation across the visual field. First, bring the target in from the periphery and ask the patient to report when the red colour is appreciated. Next, probe different regions of the field looking for relative or absolute scotomas. If a partial hemianopic defect is suspected (the patient is able to detect a moving finger or count fingers but there is colour desaturation), the target can be presented moving across the vertical meridian, where the sudden change in the appearance of the colour will be readily appreciated by the patient. Figure 1 shows some common patterns of visual field defects.

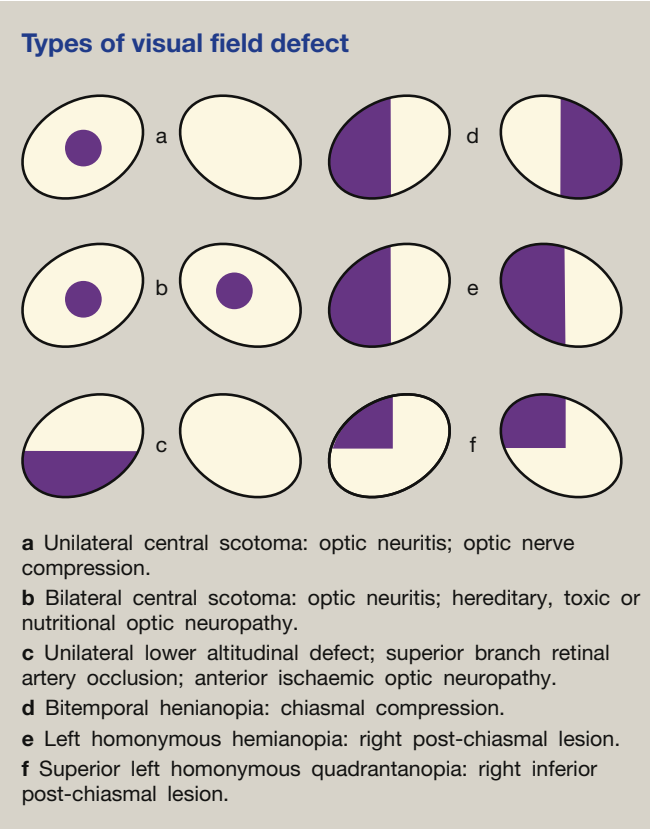

Figure 1

The pupils

The pupillary light reflex is of great importance and should be examined next. As bright a light source as possible should be used in dim surroundings. First, it must be established that there is a direct and consensual response to light in each eye. In severe visual loss, the amplitude of the pupil response to light is diminished; indeed, it will be absent if there is no perception of light in the eye (care must be taken that stray light does not fall onto the unaffected eye, producing a spurious consensual response).

If there is partial loss of vision due to unilateral optic neuropathy, the direct light reflex can appear symmetrical, but if the light is swung from one eye to the other, both pupils will dilate when the light is moved to the abnormal eye. This sign, known as the relative afferent pupillary defect (RAPD), is a sensitive test of unilateral (or asymmetrical bilateral) optic neuropathy (Figure 2). It is also reasonably specific because, as with colour vision, the pupil light reflex is affected in moderate optic neuropathy, whereas retinal disease needs to be more severe before an RAPD is detected. If both optic nerves are affected equally, it is not possible to demonstrate an RAPD. The pupil light reflex is not affected if the problem is an optical one, nor is it affected in cortical blindness.

The fundus

Examination of the fundus, using the direct ophthalmoscope, is also a very useful skill for physicians to acquire. They must become accustomed to the range of normal appearances, such as the difference between a myopic eye (a large eye with a large