have ever been in hospital or consulted a doctor. The question may need to be repeated three times, because patients may hear and answer only the first point about accidents.

Family history
- "Is there anybody in the family with anything like this?"
  The family history is often relevant in neurological disease, and may give additional information about the domestic situation that will not otherwise emerge. The author asks in detail about first-degree relatives (parents, siblings and children) and, if any of them have died, about the cause of death. If a familial condition is suspected, a detailed family tree should be constructed and the possibility of consanguinity raised.
- "Are your parents alive? How old is your father/mother? Is he/she well?"
- "Do you have any brothers or sisters?" Ask about their ages and health.
- "Do you have any children?" Ask about their gender, age and health.

Personal history: details of patients’ social, business and domestic background is often relevant in determining the management of many conditions. It is necessary to know their occupation, how they get to work, who is at home with them, what degree of support is available (psychological and physical) and their housing conditions. Sometimes these questions reveal unexpected information such as the recent arrival of a demented mother-in-law, the stress of caring for a disabled child, redundancy of the patient or partner, social isolation because of the death of the partner, stress related to illness in the partner, or the fact that the patient is a single mother without support, which would make admission to hospital difficult.

This is a good opportunity to ask patients how their condition has affected their life.
- "How much time have you missed from work because of this problem (in the last month, 6 months, year)?"
- "Does this problem ever prevent you from going out or limit your activities?"

Some patients describe severe blinding headaches, but are nevertheless capable of going out in the evening or continuing work. This gives some independent measure of the severity.

Patients with increasing neurological deficit (e.g. multiple sclerosis or Parkinson’s disease) often undertake fewer activities as their horizons shrink; this may occur as a result of increasing disability, or of frequency and urgency of micturition. These matters should all be addressed, and effective management of them may improve the patient’s quality of life more than any attempt to treat the underlying condition.

Concluding the history
Before proceeding with the examination, ask the following question.
- "Is there anything else you would like to tell me?"

Failure to take a good history is a common cause of failure to reach the correct diagnosis. Failure to explain to the patient what you think and how you intend to proceed is the most common cause of complaint, so conclude the consultation with:
- "Is there anything else you would like to ask or discuss?"

Neurological eye problems
Masud Husain
Christopher Kennard

Many neurological disorders present with visual dysfunction (e.g. loss of vision, diplopia). The history and examination (Figures 1 and 2) help to reduce the number of possible diagnoses.

Ocular vascular disease
Central retinal artery occlusion leads to sudden, usually painless, monocular blindness. When the occlusion is temporary, the patient may complain of transient monocular visual loss (amaurosis fugax, Figure 3), with blindness described as ‘coming down like a curtain’. Permanent occlusion of the central retinal artery leads to retinal infarction and blindness. When only a branch retinal artery is occluded, an altitudinal field defect may occur, with visual loss in either the upper or the lower half of the visual field. Fundoscopy may demonstrate embolic material within blood vessels, retinal oedema and, later, a cherry-red spot where the vascular choroid at the macula stands out against a pale, infarcted retina.

Insufficient blood flow in the central retinal artery (temporary or permanent) may result from thromboembolism, vasculitis, spasm or reduced perfusion (e.g. critical carotid stenosis). Examination and investigations are similar to those in transient ischaemic attacks or stroke, and the principles of treatment and prophylaxis are also the same. However, it is important to exclude intraocular pressure (e.g. glaucoma) as an underlying cause.

Anterior ischaemic optic neuropathy results from infarction of the optic nerve head caused by occlusion of the posterior ciliary arteries. Patients typically complain of sudden, painless, monocular visual loss that is usually altitudinal. The disc is swollen and commonly pale; there may be haemorrhages and cotton-wool spots. This condition is most commonly non-arteritic caused by atherosclerosis, or arteritic usually associated with temporal arthritis and raised ESR. Prompt administration of high-dose corticosteroids (see page 000) is important to prevent visual loss in the other eye when temporal arthritis is suspected.

Masud Husain is Reader in Neurology and Wellcome Trust Senior Fellow at Imperial College London and the West London Neurosciences Centre, Charing Cross Hospital, London, UK. He qualified from the University of Oxford. His research interest is disorders of visual perception in stroke and neurodegenerative conditions.

Christopher Kennard is Professor of Clinical Neurology and Deputy Principal of the Faculty of Medicine at Imperial College London and Honorary Consultant Neurologist at Charing Cross Hospital, London, UK. His principal research interest is neuro-ophthalmology, particularly disorders of eye movement in neurological disease.
Optic neuropathies

Non-vascular optic neuropathies usually present as monocular visual disturbance with central scotoma, impaired colour vision and a relative afferent pupillary defect. Causes include:

- optic neuritis
- compression of the optic nerve by retrobulbar masses (e.g. tumours, connective tissue swelling as in Graves’ disease)
- granulomatous disease (e.g. sarcoidosis)
- metabolic alterations (e.g. vitamin B₁₂ or B₁ deficiency)
- drugs or toxins (e.g. ethambutol, isoniazid, lead).

Tobacco–alcohol amblyopia is optic neuropathy caused by tobacco, alcohol or nutritional deficiencies (e.g. B₁₂, B₆, B₁).

Optic neuritis results from demyelination and is the most common cause of acute optic neuropathy. It presents as progressive visual loss over a few days, improving during subsequent weeks. It may be associated with a dull ache of the eye that is exacerbated by eye movement. Patients commonly complain that colours become drab and the world looks grey. Testing with Ishihara colour plates may reveal a defect, and red objects often appear desaturated. There may be a central scotoma and a relative afferent pupillary defect. The optic disc may be swollen (papillitis) or may appear normal (retrobulbar neuritis). There may be optic disc pallor as a result of optic atrophy. Intravenous methylprednisolone, 1 g/day for 3 days, reduces the symptoms but does not affect the outcome.

Corticosteroid treatment is indicated only when the acuity is worse than 6/36 or the pain is significant. In the UK, 75% of these patients develop multiple sclerosis.

An increasing number of unexplained cases of optic neuropathy are being identified as Leber’s hereditary optic neuropathy, a result of point mutations in the mitochondrial genome.

Optic disc swelling

Swelling of the optic disc (Figure 4) may result from local optic nerve pathology (e.g. inflammation, infiltration, meningioma). Swelling caused by raised intracranial pressure (e.g. intracranial mass, hydrocephalus, cerebral venous thrombosis, benign intracranial hypertension) is termed ‘papilloedema’. Visual acuity is reduced in optic nerve pathology but is normal in papilloedema. In papilloedema, the blind spot is enlarged because the optic nerve head swells.
Papilloedema should be distinguished from drusen (deposits of hyaline-like calcific material), which may cause elevation of the optic disc head (Figure 5), and from medullated nerve fibres, which have accompanying myelin sheaths beyond the lamina cribrosa. Benign intracranial hypertension presents with headache, swollen optic discs and raised intracranial pressure; ventricular size and CSF are normal. It usually occurs in obese young women, who may complain of transient visual obscuration. There is an association with oral contraceptives, corticosteroids and several antibiotics, including tetracycline. Treatment comprises stopping any drugs that may be responsible, therapeutic lumbar puncture (sometimes sequential) and acetazolamide, 250 mg b.d. initially, increasing to 1 g if side-effects permit, to reduce CSF production.

Chronic papilloedema leads to compressive optic neuropathy and progressive visual loss. It is therefore essential to monitor the patient’s visual fields and acuity at regular intervals. If simple measures fail, optic nerve sheath fenestration to decompress the optic nerve or peritoneal shunting may be required.

Dural sinus thrombosis may lead to a clinical syndrome similar to that of benign intracranial hypertension. It can be detected by magnetic resonance angiography or CT with contrast (Figure 6). Anticoagulation should be commenced.

Optic chiasm, tract and radiation lesions

Bitemporal (heteronymous) field defects are the hallmark of an optic chiasm lesion. The most common presentation of a chiasmal lesion is with upper quadrant defects, which suggest compression from below, usually by a pituitary adenoma. Visual acuity may be reduced when the optic nerve is involved; therefore, always check the upper temporal field in the contralateral eye when assessing apparently monocular optic neuropathy. Pressure on the chiasm from above leads initially to lower quadrant defects and suggests craniopharyngioma or third ventricular tumour. Compression may lead to optic atrophy, and occasionally papilloedema associated with suprachiasmal lesions occurs. Primary pituitary disorders or secondary compression may also lead to endocrine abnormalities (e.g. hyperprolactinaemia, diabetes insipidus).

Homonymous field defects occur with retrochiasmal lesions, usually cerebral infarcts and tumours. A field defect that is more

Causes of optic disc swelling

- Ocular disease – uveitis, vein occlusion
- Inflammatory – neuroretinitis, optic neuritis
- Vascular lesions – anterior ischaemic optic neuropathy, cranial arteritis, systemic arteritis
- Infiltrative lesions – lymphoma, reticuloendothelial
- Systemic disease – anaemia, hypoxaemia, hypertension
- Raised intracranial pressure – mass lesion, benign intracranial hypertension, hypertension
substantial in one eye (incongruous homonymous hemianopia) suggests a lesion of the optic tract. Damage to the parietal fibres of the optic radiations characteristically produces congruous inferior homonymous quadrantanopia, whereas injury to the temporal fibres leads to a congruous superior quadrant defect.

**Lesions of the cerebral cortex**

Unilateral lesions of occipital cortex lead to congruous homonymous hemianopia. When the cause is vascular, macular vision is generally spared, perhaps because of the dual blood supply (from posterior and middle cerebral arteries) to the occipital pole.

Damage to the posterior parietal cortex, particularly the right hemisphere, is often associated with disorders of visual attention, including extinction (when two stimuli are briefly presented simultaneously, the patient reports seeing only the one on the side of the lesion) or hemineglect (the patient ignores objects contralateral to the lesion, Figure 7).

Unilateral lesions of the frontal cortex may result in unopposed activity of the contralateral frontal eye field, and lead to conjugate gaze deviation towards the side of the lesion (and away from the side of any associated hemiplegia).

**Ptosis**

Weakness of the levator palpebrae superioris (innervated by the IIIrd nerve) or the smooth muscle fibres of Muller’s muscle (innervated by sympathetic fibres) leads to ptosis. There are many central and peripheral causes. An important cause of acute-onset ptosis and Horner’s syndrome is carotid artery dissection, which may occur after relatively mild neck trauma. Definitive diagnosis requires MRI or angiography.

Anticoagulation is usually recommended to reduce the risk of embolism and cerebral infarction.

**Ophthalmoplegia**

Diplopia is the most common complaint of patients with limitation of gaze.

*External ophthalmoplegia* is paralysis of the extraocular muscles. (Internal ophthalmoplegia refers to paralysis of the sphincter pupillae and ciliary muscle.) Lesions causing external ophthalmoplegia may be infranuclear (e.g. myasthenia gravis, mass lesions within the orbit or cavernous sinus or near the brain stem), internuclear (see below), nuclear (e.g. vascular disorders or demyelination affecting the brain stem nuclei of the IIIrd, IVth or VIth nerves) or supranuclear (e.g. frontal infarcts). In patients with IIIrd nerve palsies, it is important to determine whether the pupilloconstrictor parasym pathetic supply is intact. When the pupil is spared, the most likely cause is vascular disease associated with atherosclerosis, hypertension or diabetes mellitus. When the pupil is involved, the patient requires magnetic resonance or intrarterial angiography to investigate the possibility that a posterior communicating artery aneurysm is compressing the oculomotor nerve. It is important to appreciate that, in some cases, though the pupil may initially be spared, it may subsequently become affected. Isolated IIIrd nerve palsies must therefore be assessed carefully by specialists.

*Internuclear ophthalmoplegia* results from a lesion of the median longitudinal fasciculus that disconnects the VIth nerve nucleus in the pons (responsible for activating the ipsilateral lateral rectus) from the contralateral IIIrd nerve nucleus in the midbrain (which activates the medial rectus in the opposite eye). In classical right internuclear ophthalmoplegia, when the patient attempts to look to the left, abduction of the left eye is normal but the right eye is unable to adduct fully because the right medial rectus is not activated in concert. The left eye usually displays jerk nystagmus. Convergence to a near stimulus may be preserved, demonstrating that the right eye can adduct fully and that there is no paralysis of the medial rectus. The most common cause of unilateral internuclear ophthalmoplegia is a small pontine infarct. In younger patients, demyelination is more often responsible for bilateral internuclear ophthalmoplegia.

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*FURTHER READING*

