Neuro-ophthalmology for General Practitioners: A Revision

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What is neuro-ophthalmology?

The term "Neuro-ophthalmology" (眼神經科) is probably unfamiliar to most people, including a lot of doctors. In the higher surgical training curricula of the Royal College of Ophthalmologists (UK) and the Royal College of Surgeons of Edinburgh, it is one of the seven established subspecialties of ophthalmology, the others being oculoplastic; cornea; cataract and refractive surgery; glaucoma; retina and paediatric ophthalmology.

Neuro-ophthalmologists mainly deal with optic nerve disorders, abnormalities of the pupil, extraocular movement, eye lid position/movement and intracranial conditions which affect the eyes. In addition, patients with visual loss of unknown cause often end up in neuro-ophthalmology clinics. Essentially neuro-ophthalmology is the fusion between neurology and ophthalmology. In the US, neuro-ophthalmologists can either be neurology-or ophthalmology-trained. Currently neuro-ophthalmology services are offered at some Hospital Authority eye clinics and patients are usually referred from the general eye clinic of the respective hospitals, like other subspeciality services.

Relevance of Neuro-ophthalmology to the General Practitioner

Although GPs are not expected to manage neuro-ophthalmological conditions by themselves, they have an important role to play as the gatekeeper: by making a basic assessment and referring appropriately, they can potentially not only save the sight but also the life of a patient.

The diagnosis of a lot of neuro-ophthalmological conditions can be made by taking a good clinical history. Often all the equipment required to make a basic assessment in a busy GP clinic are a pocket Snellen chart, pen torch and an ophthalmoscope (see Table 1). We will see how this is applied to specific clinical situations below.

Table 1: The Basic Neuro-ophthalmological Assessment

- History
- Inspection
- Examination (directed)
  - Visual acuity
  - Pupils
  - Extra-ocular movement
  - Visual field
  - Fundus
- Other associated signs (other cranial nerves)

Specific clinical situations:

A. Visual loss

In the history, ascertain whether the visual loss is in one or both eyes, the onset and duration; and whether there are any associated symptoms. Patients are often unable to distinguish between visual loss in e.g. in the right eye or the right side of visual field of both eyes. One should therefore ask them to cover either eye to see if the visual problem persists. Past medical history, particularly diabetes and hypertension, is relevant, since they can cause diabetic/hypertensive retinopathies and are risk factors for e.g. retinal artery/ vein occlusion and non-arteritic anterior ischaemic optic neuropathies (the risk of developing these conditions is further increased by smoking). A drug history may be contributory: it is well recognised that ethambutol and isoniazid can cause optic neuropathy and chloroquine/ hydroxychloroquine can cause retinopathy. More recently it has been identified that erectile dysfunction drugs and amiodarone may also be associated with optic neuropathies and vigabatrin can lead to visual field defects.

In the examination, apart from testing the visual acuity of each eye (with appropriate glasses and pinhole if necessary), the pupillary light reflex is very important as it is one of the only objective signs of optic nerve dysfunction. There is no relative afferent defect if the visual loss is bilateral and equal; and cortical visual loss (posterior to the midbrain in the visual pathway) does not affect the pupillary reaction. Testing of visual fields is particular relevant if the patient complains of bilateral visual loss.

If the patient has unilateral visual loss, it is most appropriate to refer to the ophthalmologist, and urgent
referral is warranted if e.g. retinal detachment or temporal arteritis is suspected. However if the patient has homonymous visual field defects in both eyes, particularly if there are other neurological deficits, referral to a neurologist or an accident and emergency department may be appropriate, depending on the clinical situation.

B. Extraocular movement abnormalities

These often manifest as diplopia. It is a priority to ascertain whether the diplopia is monocular or binocular: ask the patient to cover up either eye and see if the double vision persists. If it does, this is monocular diplopia, most likely caused by a problem with the optic media in the affected eye and referral to an ophthalmologist is warranted. If the diplopia is binocular then use the approach in Table 2 to further narrow down the diagnosis.

The effects on extraocular movement resulting from third (oculomotor), fourth (trochlear) and sixth (abducens) nerve palsies would be well known to the reader. In terms of symptoms, sixth nerve palsy results in vertical or oblique diplopia. Traditionally, pressure can cause sixth nerve palsy without a lesion along the nerve itself. Third and fourth nerve palsies also be a false localising sign i.e. increased intracranial pressure can cause sixth nerve palsy without a lesion along the nerve itself. Third and fourth nerve palsies will result in vertical or oblique diplopia. Traditionally, third nerve palsies are divided into ‘medical’ or ‘surgical’, depending on whether the pupil is involved. If it does, this is monocular diplopia, most likely caused by a problem with the optic media in the affected eye and referral to an ophthalmologist is warranted. If the diplopia is binocular then use the approach in Table 2 to further narrow down the diagnosis.

<table>
<thead>
<tr>
<th>Table 2: Clinical Approach to Diplopia</th>
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<tbody>
<tr>
<td><strong>History</strong></td>
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<td>Monocular or binocular?</td>
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<td>Vertical or horizontal?</td>
</tr>
<tr>
<td>Onset and duration</td>
</tr>
<tr>
<td>Variable?</td>
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<tr>
<td>Associated problems e.g. ptosis?</td>
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<tr>
<td><strong>Examination</strong></td>
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<tr>
<td>Inspection (squint?)</td>
</tr>
<tr>
<td>Cranial nerve pattern or not?</td>
</tr>
<tr>
<td>Lids and pupils</td>
</tr>
<tr>
<td>Other cranial nerve functions:5, 7, 8</td>
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</table>

If the extraocular movement abnormalities fall into a cranial nerve pattern, it is important to check for other associated neurological defects, which can pinpoint the location of the lesion (see Table 3).

<table>
<thead>
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<th>Table 3: Localization of lesions causing 3rd/ 4th and 6th nerve palsies</th>
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<tr>
<td>Neurological signs</td>
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<tr>
<td>Cranial nerves 2, 3, 4 and 6 palsies + proptosis</td>
</tr>
<tr>
<td>Cranial nerves 3, 4, 5 (ophthalmic division) and 6 palsies</td>
</tr>
<tr>
<td>Cranial nerves 5, 6, 7 &amp; 8 palsies + cerebellar</td>
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If the extraocular movement defects do not fall into a cranial nerve pattern, it may be caused by thyroid eye disease (most common), myasthenia gravis or orbital lesions. Thyroid eye disease (ophthalmic Graves) can occur not only in the thyrotoxic state, it can also occur in patients who are euthyroid or hypothyroid. Thyroid eye disease can often be diagnosed clinically and if necessary, confirmed by the typical appearance of enlarged extraocular muscles with sparing of muscle tendons on computer tomography of the orbit. In recent years, smoking has been identified to be the most important risk factor for developing eye disease in patients with Graves disease.

Ocular myasthenia gravis (OMG) will result in variable diplopia and ptosis and the pupil is never involved. Traditionally OMG can be diagnosed with the Tensilon test. However, more recently the ‘Ice pack’ test has largely replaced Tensilon test for the clinical diagnosis of OMG in patients with ptosis. It involves placing an ice pack on the ptotic eye for 2 minutes which should improve the ptosis. It is easier and safer to perform than the Tensilon test, has a high specificity and sensitivity, but it is of limited use in patients with only subtle extraocular movement limitation without ptosis.

C. Unequal sized pupils (Anisocoria)

The pupil size is controlled by the sympathetic and parasympathetic nervous systems. Disruption of the sympathetic pathway will result in Horner’s syndrome, which manifests as a small, poorly reactive pupil (miosis), mild ptosis +/- lack of sweating on the ipsilateral side of the face (anhidrosis). Disruption of the parasympathetic pathway will result in an enlarged pupil, e.g. in pupil-involving oculomotor nerve palsy or in Adie’s syndrome. Adie’s syndrome (also called Homans-Adie tonic pupil syndrome) is a condition which typically affects young adult females, unilateral in 80% of the cases, and manifests as a large pupil, which is poorly reactive to light, but reacts to accommodation (“light- near dissociation”). This condition may be associated with hyporeflexia or areflexia.

The most common causes of unequal sized pupils seen by ophthalmologists are local causes at the iris itself—e.g. traumatic/iatrogenic damage to the iris, dilating eye drops, glaucoma eye drops (pilocarpine), acute iritis etc. In particular, a red painful eye, associated with headache, nausea, hazy cornea and a poorly reactive, semi-dilated pupil should suggest acute angle closure glaucoma and warrants urgent referral to an ophthalmologist or an accident and emergency department.

D. Ptosis

The classification of the causes of ptosis or droopy eyelids can be simplified into 1) muscular/mechanical, 2) neurological (third nerve palsy and Horner’s syndrome) and 3) neuro-muscular (myasthenia gravis). When seeing a patient with ptosis, the following aspects of the history need to be ascertained:

a) Whether the ptosis is unilateral or bilateral
b) The duration—old photos including the photo on the HKID card, give a good clue
c) Whether it is associated with abnormalities of the pupil and extraocular movement; and
d) Whether it is variable, which might suggest myasthenia gravis

The most common cause of ptosis in the elderly is aponeurotic ptosis, which results from dehiscence, disinsertion or stretching of the levator aponeurosis. It is usually insidious in onset, bilateral, and affected patients will have widened upper lid creases, deep superior sulci with good preservation of levator...
function. We also have to be aware that local causes, e.g. severe conjunctivitis can cause lid swelling and mild ptosis in the acute stage, as can a retained contact lens.

E. Optic disc abnormality
In practice, it is difficult to get a clear view of the optic discs through undilated pupils and with a direct ophthalmoscope, at it is the case in the GP’s office or at the accident and emergency department.

‘Papilloedema’ is a term reserved for bilateral optic disc swelling secondary to intracranial pressure. Although it may be associated with transient visual obscurations, the vision usually remains normal until the late stage and there may be other signs or symptoms of increased intracranial pressure.

Optic disc swelling due to all other causes are simply described as 'disc swelling', even when it is bilateral. Disc swelling can be 'real' and pathological, for example in anterior ischaemic optic neuropathy, optic nerve compression, malignant hypertension or optic neuritis. However it is to be noted in typical idiopathic optic neuritis and that associated with multiple sclerosis, about two thirds are ‘retrobulbar’ in nature i.e. although the patient complains of visual loss with objective visual dysfunction (i.e. relative afferent pupillary defect), the disc appearance can be entirely normal- to put it succinctly, it is a condition in which ‘the patient sees nothing and the doctor sees nothing’.

There are some patients with physiologically elevated discs, which is termed ‘pseudo- disc swelling’. This can occur in patients with hypermetropia, tilted discs or patients with optic nerve head drusen. Usually these patients are asymptomatic and the ‘disc swellings’ are incidental findings. Distinguishing between real and pseudo-disc swelling sometimes pose difficulties even for an experienced ophthalmologist, therefore general practitioners are certainly not expected to make the distinction and referrals to ophthalmologists are warranted if optic disc swelling is suspected.

### Urgent neuro-ophthalmological conditions

Of particular importance to the general practitioner are urgent neuro-ophthalmological conditions which warrant prompt referral and treatment. (Table 4)

#### Table 4: Urgent neuro-ophthalmological conditions

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<tr>
<th>Conditions</th>
<th>Clues to diagnosis</th>
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<tr>
<td>Temporal arteritis</td>
<td>Elderly, general malaise, weight loss, Headache, scalp tenderness, jaw claudication, Tender and pulsatile temporal arteries, Visual disturbance (transient or persistent), Raised ESR and CRP.</td>
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<tr>
<td>Intracranial aneurysm (Especially of posterior communicating artery)</td>
<td>Surgical 3rd nerve palsy; incomplete/ pupil involving/ headache; Atypical age group and no vasculopathic risk factors.</td>
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<tr>
<td>Carotid dissection</td>
<td>Acute onset Horner’s syndrome, Painful neck, (Vertebral dissection can also cause Horner’s syndrome, but maybe associated with other signs of lateral medullary syndrome).</td>
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</table>

### Conclusion

A lot of neuro-ophthalmological diagnoses can be made by taking a careful history and by performing a directed examination using basic tools available at the GP’s office. The general practitioner can really make a difference by making appropriate referrals after a preliminary assessment. Save sight, save life and impress your colleagues with your diagnostic skills!

### References