Double vision

assessment and initial management

Double vision is an important symptom with a large number of possible causes. An accurate initial assessment is extremely helpful in assessing the urgency of the situation and determining the appropriate course of action.

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Double vision (diplopia) is a common neurological symptom, but it can be a daunting one for the assessing GP because the list of potential causes is enormous. However, by following a few simple guidelines, it is possible to diagnose the more common causes with confidence. This article aims to provide a straightforward guide to assessment, along with advice on the initial management of the more common conditions.

Examination of the patient with diplopia is much easier when the relevant anatomy is understood and appropriately applied. A practical summary of the structure and function of the extraocular muscles is provided in the box on page 28, and a list of important causes is given in Table 1.

History

As always in neurological diagnosis, the first thing to try to determine is the site of the lesion.

The diplopia itself

If the patient is a good witness, asking whether the double vision disappears when either eye is covered will confirm whether the problem is binocular (arising from malalignment of the eyes) or monocular. However, it is not always possible to obtain this information.

The nature of the image separation often gives a clue as to the site of the lesion. Horizontally separated images are most often due to a 6th (abducens) nerve palsy, although there are many differential diagnoses. Vertical separation has a

IN SUMMARY

- **Monocular diplopia should be detected at an early stage because it is managed differently to binocular diplopia, with patients requiring ophthalmological (rather than neurological) referral.**
- **Eye movements should be assessed in an H-pattern.**
- **Diplopia is greatest when the patient looks into the field of action of the weak muscle (or muscles), with the most eccentric of the two images coming from the paretic eye.**
- **Straightforward palsies of the 3rd, 4th and 6th cranial nerves can usually be diagnosed clinically.**
- **Painful ocular motor nerve palsies should be treated as an emergency and referred immediately to exclude the possibility of intracranial aneurysm. This also applies to a sudden onset, pupil-involving third nerve palsy.**
- **If symptoms fluctuate over time, consider myasthenia gravis.**
- **If there is a history of trauma, consider an orbital blowout fracture and refer the patient immediately to an ophthalmologist.**
- **Patients with more complicated disorders will need formal neurological assessment.**

Understanding the control of eye movement

The extraocular muscles

There are six muscles that move each eye. Essentially, the eyeball rests in the orbit on a hammock of fibrous tissue. It can therefore rotate in three planes: horizontal, vertical and torsional.

It is important to realise that the action of any muscle at a given point in time is determined by the starting position of the eyeball. For example, the superior oblique is almost a pure depressor (i.e. it moves the eye down) if the eye starts from an adducted (looking in) position, but it is a pure intorter of the abducted (looking out) eye.

The assessment is simplified enormously by recalling three facts:

- the horizontal muscles are conveniently placed for moving the eye horizontally from the primary position (Figure 1a)
- the superior and inferior recti are aligned with the orbital axis, which runs about 22.5° lateral to the primary position (Figure 1b)
- the oblique muscles approach the eyeball from the front and medial side (Figure 1c).

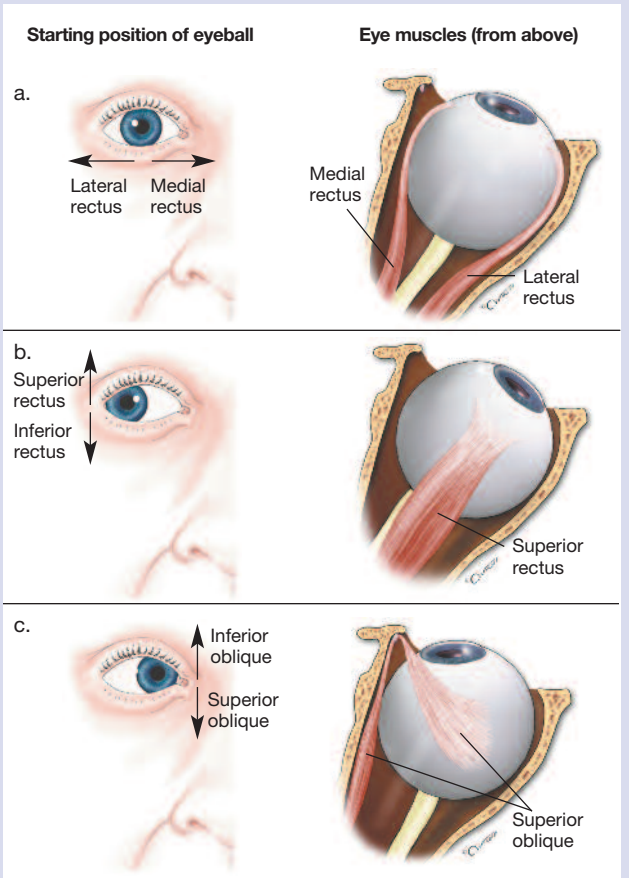

Figure 1. The action of the eye muscles depends on the starting position of the eyeball. a (top). If the eye is in the primary position, the medial rectus adducts it and the lateral rectus abducts it. b (middle). If the eye is first abducted, the superior rectus elevates it and the inferior rectus depresses it (inferior rectus not shown). c (bottom). If the eye is first adducted, the superior oblique depresses it and the inferior oblique elevates it (inferior oblique not shown).

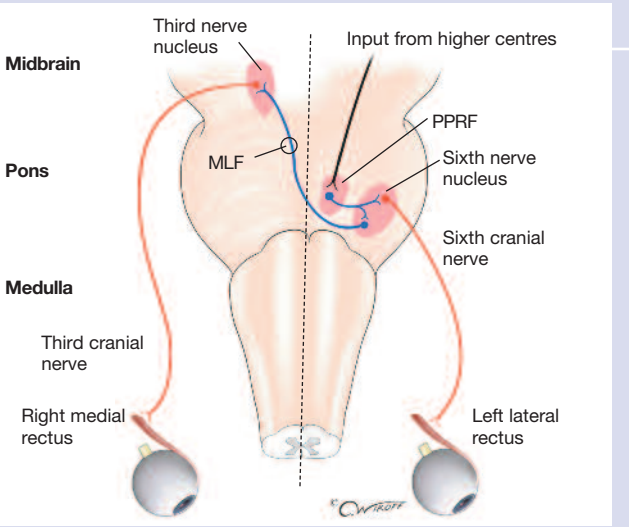

Figure 2. Higher control of horizontal eye movements. Descending inputs reach the sixth nerve nucleus via the paramedian pontine reticular formation (PPRF). There are two outputs from the sixth nerve nucleus: one directly to the ipsilateral lateral rectus via the sixth nerve, and the other to the contralateral medial rectus via the medial longitudinal fasciculus (MLF) and third nerve.

Nuclear and infranuclear connections

Three nerves supply the extraocular muscles:

- the lateral rectus is supplied by the 6th cranial (abducens) nerve.
- the superior oblique is supplied by the 4th cranial (trochlear) nerve.
- the remaining four muscles, the levator of the eyelid and the pupil are supplied by the 3rd cranial (oculomotor) nerve.

All nerves arise from their respective nuclei, travel through the brainstem, subarachnoid space, cavernous sinus, superior orbital fissure and then the orbit to reach their respective muscles.

Supranuclear connections

The co-ordination of eye movements is achieved in the brainstem. Horizontal eye movements are co-ordinated in the pons (Figure 2). Vertical and torsional eye movements as well as convergence are largely co-ordinated in the midbrain.

Internuclear ophthalmoplegia occurs as a result of a lesion of the MLF (Figure 3). There may be associated nystagmus of the unaffected eye.

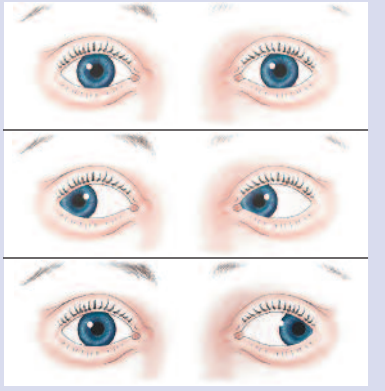

Figure 3. Right sided internuclear ophthalmoplegia due to a lesion of the right medial longitudinal fasciculus. From the primary position (top), both eyes move normally to the right (middle), but the right eye does not adduct properly on attempting to look to the left (bottom).

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large number of potential causes, but 4th (trochlear) nerve palsy, 3rd (oculomotor) nerve palsy and myasthenia gravis are high on the list of possibilities. If there is torsional separation (i.e. the images are tilted with respect to each other), a 4th nerve palsy is usually the cause.

A good witness may be able to indicate the direction of gaze that produces the greatest separation of the images, and this will narrow down the possible causes. For example, diplopia that is worst when looking horizontally to the right is likely to be due to either a right 6th nerve palsy or left medial rectus palsy. Unfortunately, however, patients are not always able to provide this information reliably, and it should always be confirmed on examination.

Timing

The timing of onset may give a clue to the nature of the pathology. Sudden onset of marked diplopia suggests the possibility of a vascular lesion, such as an infarct of one of the ocular motor nerves (i.e. the 3rd, 4th or 6th), a subarachnoid haemorrhage or stroke. Pain associated with an ocular motor nerve palsy should be treated as being caused by an aneurysm until proven otherwise; such patients require referral for immediate neurosurgical assessment, even though most will turn out to have nerve infarcts.

Onset with gradual worsening could be due to a number of different causes, including myasthenia gravis, thyroid ophthalmopathy, space occupying lesions (in the orbit, cavernous sinus or intracranially), inflammatory disease (e.g. multiple sclerosis) or, rarely, nutritional disorders (e.g. Wernicke's encephalopathy due to thiamine deficiency). If the degree of double vision varies over time, myasthenia gravis should be strongly considered.

Orbital fractures can cause diplopia due to tethering of extraocular muscles, and a fracture can sometimes be missed on skull x-ray. The best outcomes result from early management, so immediate referral to an ophthalmologist is recommended for a patient complaining of double vision after a facial or head injury that might have resulted in an orbital fracture.

Other features

There are, of course, features in the general history that may give a clue to the diagnosis. For example, a patient may have diabetes (which would suggest a vascular lesion), features of thyroid disease, or multiple sclerosis. Alternatively, there may be weakness elsewhere suggesting, for example, myasthenia gravis or stroke.

Examination

For the purposes of evaluating a complaint of double vision, the following steps are important. At the end of the examination, it should be possible to confirm or refute four 'straightforward' diagnoses: monocular diplopia, and isolated palsies of the 3rd, 4th or 6th cranial nerve. Details of the expected findings are given in Table 2.

Assess vision and examine fundi

For each eye, check visual acuity, perform a confrontational visual field test and examine the fundus. These may give a clue to the presence of refractive or retinal abnormalities that might explain monocular diplopia. Longstanding amblyopia is often associated with a congenital squint (strabismus). Unilateral visual field abnormalities very occasionally give rise to diplopia.

Check head position and other general physical signs

Check for head tilt (i.e. the head not being held vertical when looking straight ahead), which is an easily missed sign. Look for orbital, eyeball, eyelid and pupillary abnormalities, and listen for an orbital bruit.

A fourth nerve palsy is often associated

Table 1. Common causes of double vision: an anatomical approach

Eye (monocular diplopia)
Disorders of the cornea, iris, lens or vitreous
Disease of the retina (especially macula)

Orbit
Damage to nerves or muscles caused by trauma, inflammation or pressure from a neighbouring lesion

Extraocular muscles
Thyroid ophthalmopathy
Myopathy
Ophthalmic surgery, as well as treated longstanding squint (strabismus)
Eye movement disorders (e.g. nystagmus)

Neuromuscular junction
Myasthenia gravis
Botulism

3rd, 4th and 6th cranial nerves
Extraneuronal pathology, which can damage the nerve at any point, including the brainstem, subarachnoid space (e.g. infection, haemorrhage), cavernous sinus (e.g. aneurysm, infection, thrombosis, tumour) and orbit
Direct nerve damage due to infarction, vasculitis, herpes zoster, nutritional disorders, Guillain–Barré syndrome, drugs

Brainstem
Stroke and other intracranial lesions – e.g. vasculitis or arteriovenous malformation, tumour, trauma, multiple sclerosis (i.e. internuclear ophthalmoplegia)
Encephalitis
Paraneoplastic syndromes
Neurodegenerative syndromes

Cerebral hemisphere
Migraine
Epilepsy
Intracranial lesions: tumour, stroke, multiple sclerosis, trauma

Other
Psychogenic/factitious diplopia

with a compensatory head tilt (i.e. a tilt towards the side opposite the lesion). Complete third nerve palsies are often (but not always) associated with complete ptosis and a dilated pupil. Thyroid ophthalmopathy may be associated with lid retraction or lid lag. Myasthenia gravis is often associated with ptosis, which may be variable or fatigable. Proptosis suggests thyroid disease or a local orbital lesion (e.g. inflammation or tumour). An orbital bruit may be heard as a result of a carotid cavernous fistula.

Assess ocular alignment in primary position

Assess ocular alignment when the patient is attempting to look straight ahead. Malalignment may be obvious to the naked eye, but small amounts can cause marked diplopia without being visible to an external observer. Ask the patient if he or she sees double.

As an aside, it is worth pointing out that a small number of conditions can cause manifestly malaligned eyes without double vision in alert patients. These are:

- monocular or binocular visual loss
- congenital strabismus (squint)
- convergence spasm (functional)
- chronic progressive external ophthalmoplegia – mitochondrial disease (e.g. Kearns–Sayre syndrome), neurodegenerative syndromes (e.g. progressive supranuclear palsy) and dystrophia myotonica
- Duane's retraction syndrome
- a glass eye.

Perform cover tests

Double vision that disappears on covering one eye but not the other is monocular (occurrence in both eyes does occur, but is uncommon). Consider use of a pinhole, which often abolishes monocular diplopia.

Double vision that disappears on covering one or other eye is binocular. Note that the eyes may move during alternating cover, with the uncovered eye taking up fixation. This movement provides useful objective evidence of the type and extent of malalignment (see Figures 4a and b).

Assess eye movements

A step-by-step guide to examination is outlined in the box on page 32. The Bielschowsky head tilt test should be performed if there is a suggestion of superior oblique weakness. Tilting the head to the side of the lesion should exacerbate the vertical separation of the eyes (Figure 5).

Interested readers are also referred to an interactive eye simulator website hosted by the University of California Davis' School of Medicine (http://cim.ucdavis.edu/eyes/eyesim.htm).

Initial management

Monocular diplopia

A patient who has monocular diplopia should initially be referred to an ophthalmologist to exclude a refractive or

Table 2. Clinical features of four simple diagnoses

	Diplopia	Eyelid	Pupil	Cover test	Eye movements
Monocular diplopia	Variable, often improved by use of pinhole	Normal	Normal, although there may be evidence of a disorder of refractive media	Diplopia relieved by covering affected eye only	Normal
3rd (oculomotor) nerve palsy	Vertical and/or horizontal	Normal, or partial or complete ptosis	Normal or dilated	Diplopia relieved by covering either eye, exotropia	Position of affected eye often down and out; limited adduction, elevation and depression
4th (trochlear) nerve palsy	Vertical with tilted images, worse looking down to side opposite lesion	Normal	Normal	Diplopia relieved by covering either eye; Position of affected eye may be higher	Resting head tilt to opposite side; tilting head to same side worsens symptoms; limited downgaze on adduction
6th (abducens) nerve palsy	Horizontal, worse looking to side of lesion	Normal	Normal	Diplopia relieved by covering either eye, esotropia	Limited abduction

retinal disorder. If no apparent cause can be found, referral to a neurologist will become appropriate to exclude a neurological cause, although this is relatively unusual. If no obvious neurological cause is found, a functional disorder must be considered.

Isolated cranial nerve palsies

A patient with an isolated ocular motor (i.e. 3rd, 4th or 6th cranial) nerve palsy needs referral. A palsy whose onset is associated with pain must be assumed to be due to an intracranial aneurysm until proven otherwise. This also applies to a painless pupil-involving third nerve palsy. Immediate referral to a neurosurgeon or hospital emergency department is mandatory in these situations. Other isolated cranial nerve palsies should be discussed with a neurologist, although isolated pupil-sparing 3rd and 6th nerve

A procedure for testing eye movements

Eye movements should be assessed in an H-pattern. Use a target, such as a finger or a pen, initially held about one metre directly in front of the patient. This distance will prevent convergence from complicating interpretation of the observations.

Remember that the false image is always the one furthest from the primary position. It is the relative spatial position of the targets that is important, not the clarity of the image. For example, it may be that the normally moving 'strong' eye has a cataract or refractive error that will degrade the image from this eye while the image from the 'weak' (paretic) eye appears sharper or more distinct.

Horizontal movements

Ask the patient to follow the target about half to one metre (25° to 45°) to their right. Double vision that worsens is due to a weak right lateral rectus or left medial rectus.

Cover one eye and then the other. This should allow an articulate patient to determine the eye that is generating the image furthest from the midline – it is this eye that is the paretic one. If the patient is not a reliable witness, look to see how the eyes move as you alternately cover one eye and then the other (see Figures 4a and b). If the eyes are too divergent (exotropic), the problem lies with the medial rectus. If they are too convergent (esotropic), the lateral rectus is at fault. Return the target to the primary position.

Repeat the procedure on the other side by asking the patient to follow the target to the left. This will assess the left lateral rectus and the right medial rectus.

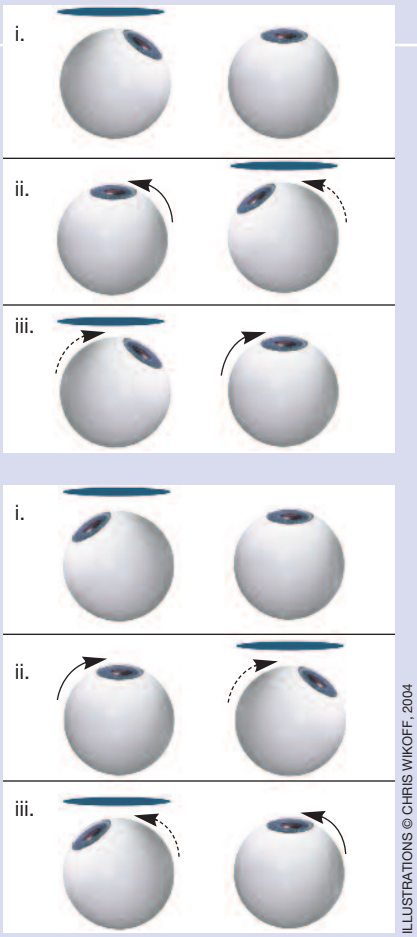
Vertical movements

The procedure for testing vertical movements is similar to that for horizontal movements, but it requires that the patient look to one side before moving the eyes up and down. This has the effect of simplifying the muscle involvement.

Ask the patient to follow the target horizontally to the right about half a metre (25°), then up about half a metre (25°). Double vision that appears or worsens could be due to weakness of the right superior rectus or of the left inferior oblique. Covering the eyes alternately should allow the patient to determine the eye that generates the higher image – this is the paretic eye. If not, covering the eyes alternately will usually help the examiner to determine the eye that is not looking up adequately.

Return the target to the horizontal baseline, then move it about half a metre vertically downwards (25°). This tests the right inferior rectus and the left superior oblique. The weaker eye will generate a lower image than the normal eye. Return the target to the primary position.

Repeat the procedure on the left side. This will test the four remaining muscles that control vertical movement (i.e. left superior rectus, right inferior oblique, left inferior rectus and right superior oblique).



Figures 4a and b. The alternate cover test. Start with one eye covered (i), then move the cover to the other eye (ii) and then back to the first eye (iii). Note whether there is any movement of the eyes as the cover is transferred. a (top panels). In esotropia, the uncovered eye moves out, implying that the eyes are too convergent. b (bottom panels). In exotropia, the uncovered eye moves in, implying that the eyes are too divergent.

The Bielschowsky head tilt test

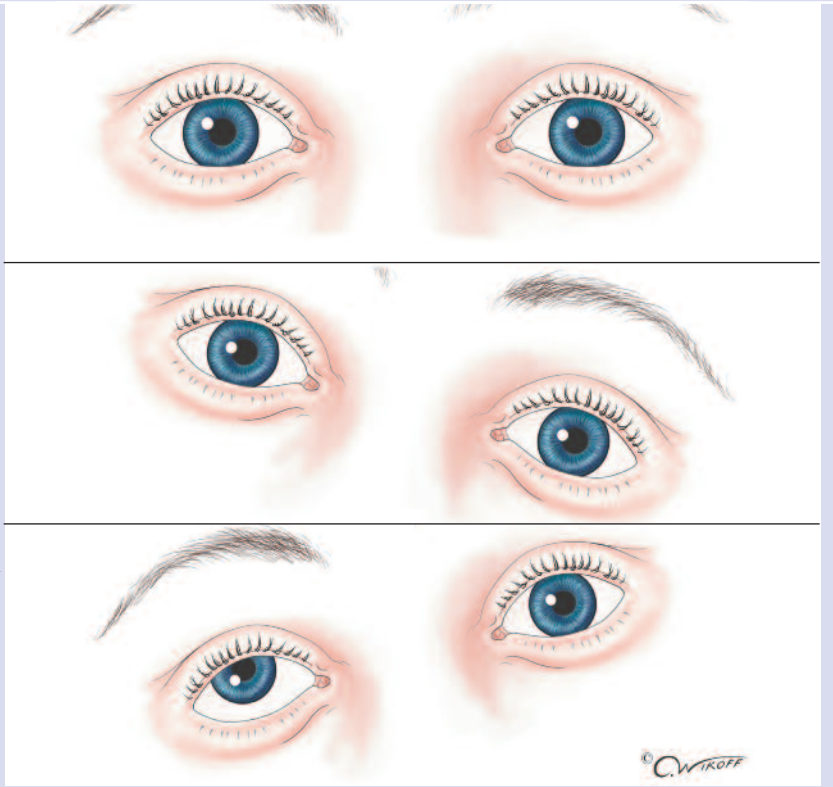


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Figure 5. The Bielschowsky head tilt test, shown here for right superior oblique palsy. Start with the eyes in the primary position (top) and tilt the head to each side (middle and bottom). When the head is tilted to the side of the lesion, vertical separation is detectable.

Consultant's comment

As the author of this article points out, diplopia can be a daunting problem because the list of possible causes is enormous. It seems likely that if 10 neurologists and ophthalmologists were asked to write such an article then there would be 10 radically differing approaches to the problem.

Associate Professor Lueck has done well in his anatomical approach and outlines the methods of examination and their significance well. I believe that the discussion of diagnosis is best considered on a similar anatomical basis from brain, midbrain, pons, oculomotor nerves and orbital structures, as he has done here.

I can commend this article for study to those wishing more information about this confusing subject.

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palsies in patients over 60 years can often be managed conservatively.

A point to note – especially for rural GPs – is that vascular 3rd and 6th nerve palsies are common in patients over 60 years of age. Therefore, if a patient presents with a painless, pupil-sparing 3rd nerve palsy or a painless 6th nerve palsy, it is reasonable to check blood pressure, ESR and blood sugar level, and to then observe (assuming these are normal). If the problem settles within two months, no further action is needed (other than attention to vascular risk factors). If there is no recovery, referral is appropriate.

Other conditions

If the diagnosis does not fit any of the above categories, the double vision should be regarded as 'complex' and the patient referred to a neurologist. One important condition to consider in this circumstance is myasthenia gravis. Clues to this diagnosis are diurnal variability and fatigability of symptoms, ptosis, weakness of other muscles, and the presence of other autoimmune conditions.

Conclusion

Double vision is an important symptom with a large number of possible causes. Accurate initial assessment of the physical signs – particularly the eye movements – is extremely helpful in assessing the urgency of the situation and in determining the appropriate course of action. MT

Further reading

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2. Borchert MS. Principles and techniques of the examination of ocular motility and alignment. In: Miller NR, Newman NJ, eds. Walsh and Hoyt's Clinical neuro-ophthalmology, vol. 1. 5th ed. Baltimore: Williams & Wilkins; 1998. p. 1169-1188.

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