

# The differential diagnosis of diplopia

Every clinician should be aware of the importance of an appropriate response to the symptom of diplopia. The aetiology can vary from uncorrected astigmatism to life threatening intracranial anomalies. A previous article (CPD Module 2 Part 6) has described many of the different causes of diplopia. This article discusses the ways in which they can be differentiated.

A review of patients presenting at a hospital casualty department complaining of diplopia found that 25% presented with monocular diplopia (Morris 1991). Of the remainder with binocular diplopia, 39% were due to infranuclear palsies, 26% were due to mechanical anomalies (muscular or traumatic), 14% were longstanding concomitant deviations or convergence/accommodation deficits, 8% were due to supranuclear palsies, 3% were due to spectacle intolerance, and in 10% of cases a cause could not be established. This paper emphasised the importance of not jumping to the conclusion that every patient complaining of diplopia has an underlying sinister pathology. In addition, as less than half the incomitancies were due to a single muscle infranuclear palsy, caution should be exercised when trying to describe an ocular motility pattern in terms of a single muscle defect.

## The major tools used in the differential diagnosis of diplopia are:

1. History and symptoms
  - Previous ocular history
  - General health
  - Family history
2. Observations
  - Abnormal head posture
  - Lid position
  - Prescription
3. Monocular tests
  - Monocular occlusion
  - Visual acuity (VA) with and without pinhole
  - Amsler chart
4. Cover Test
5. Ocular Motility
6. Near point of convergence
7. Fixation disparity
8. Measurement of the angle of deviation
  - Prism cover test
  - Maddox double rod test
9. Bielschowsky
10. Fusional reserves
11. Past pointing
12. Diplopia chart
13. Field of BSV
14. Hess chart

On presentation of any acute onset ocular motor deficit, one of the most difficult but important decisions to make is whether it is of recent onset or due to the decompensation of a long standing deviation. Some of the best clues can be obtained from the patient's response to specific questions. Most patients with an acute onset condition know exactly when the diplopia started (to the minute), the direction of gaze affected and the direction of separation of the images. If they have an abnormal head posture (AHP), they often have an associated neck ache. They normally attend for an eye examination within one to two days of the onset of symptoms. In contrast, patients with long standing deviations, normally find it difficult to be specific about the time of onset of the diplopia and the description of the image separation is more vague. If such a patient has an AHP, they may find it uncomfortable to straighten their head.

Any patient presenting with diplopia should be examined carefully to exclude the possibility of sinister aetiology. Neurological disease affecting the ocular motor system most commonly results in an incomitancy, but can present as a concomitant deviation (Hoyt and Good, 1995). Suggested below is a diagnostic routine and methodology that can be used in the event of a patient presenting with diplopia. Particular emphasis is placed on techniques that can be used for differential diagnosis. This is followed by a summary of many of the different classes of defect that can lead to diplopia, and their specific differential characteristics.

## Diagnostic techniques

### 1. History and symptoms

The value of a good history should never be underestimated in the differential diagnosis of diplopia. Careful description of the diplopic images can help differentiate monocular or refractive diplopia from a problem of the ocular motor system, the former often being describe as a 'ghosting' rather than a true diplopia. The frequency with which diplopia is recognised can provide useful information. A patient who has suddenly recognised physiological diplopia will generally report it as being present only when they are tired and then only fleetingly. Questioning about the diplopia can indicate the acuteness of the onset, as can clues from the previous ocular and medical history. Other ocular symptoms

can also be a useful diagnostic tool such as partial ptosis in a IIIrd nerve palsy. Monocular eye lid closure in sunlight can indicate either photophobia (Wiggins and von Noorden, 1990) or diplopia due to a distance exotropia (Wang and Chryssanthou, 1988). Ocular symptoms in dysthyroid eye disease include 'gritty eyes', lid retraction, chemosis in the orbital region and proptosis. Dysthyroid eye disease is normally, but not exclusively, related to hyperthyroidism (Fells, 1991).

### Previous ocular history

An AHP which has been present since childhood indicates a congenital incomitancy. History of occlusion therapy or childhood surgery may indicate a recent decompensation of a long-standing deviation or a breakdown in suppression. History of eye exercises or use of prismatic lenses points to the previous decompensation or intermittent binocular control of a long-standing deviation.

### General health

The general health or well-being of a subject may reveal signs associated with a systemic condition such as the weakness and fatigue associated with Myasthenia Gravis, or previous episodes of poor vision that could be associated with Multiple Sclerosis. Vertigo and tinnitus are symptoms of a middle or inner ear defect, which can occasionally be associated with a VIth nerve lesion (Gradenigo's syndrome). Equally, headaches can be associated with diplopia, either in relation to asthenopic symptoms (they get worse after prolonged close work), ophthalmic migraine or intra-cranial anomalies.

### Family history

Family medical and ocular history can also be of diagnostic value in long-standing and recent onset conditions, especially where there is family history of hypermetropia or an ocular motor disorder.

### 2. Observations

#### Abnormal head posture

An AHP developed to compensate for an incomitancy is usually adopted to move the eyes into a position of comfortable binocular vision, in which case it can be a useful diagnostic tool. The presence of an AHP normally indicates that the patient has the capacity for good binocular functions. Using an AHP to move the eyes out of the direction

of action of the defective muscle effectively makes the chin 'point' in the direction of action of the defective muscle. Normally, the right superior oblique (SO) moves the right eye down and to the left, so the AHP would be chin down and to the left, with a head tilt generally away from the affected side (although this can be inconsistent). The 'text book' AHP for each single muscle palsy is shown in **Table 1**, but this can be misleading, as moving the eyes out of the direction of action of a single muscle palsy is not the only reason for developing an AHP.

An AHP could also:

- put the eyes into a position that will bring the visual axis of one eye into a suppression scotoma.
- separate the images to minimise the effect of confusion.
- move the eyes out of the direction of a mechanical limitation of eye movement.
- improve visual acuity in patients with ptosis or nystagmus.
- be non-ocular, such as in torticollis (spastic contraction of a neck muscle).

**Table 1**

The 'text book' abnormal head posture shown for each individual extra-ocular muscle

Defective muscle	Affecting right eye			Affecting left eye		
	Chin	Turn	Tilt	Chin	Turn	Tilt
Lateral Rectus (LR)	X	Right	X	X	Left	X
Medial Rectus (MR)	X	Left	X	X	Right	X
Superior Rectus (SR)	Up	Right	Right	Up	Left	Left
Inferior Rectus (IR)	Down	Right	Left	Down	Left	Right
Superior Oblique (SO)	Down	Left	Left	Down	Right	Right
Inferior Oblique (IO)	Up	Left	Right	Up	Right	Left

**Lid position**

Unilateral partial ptosis will often accompany a SR paresis, as the SR and levator palpebrae superioris are innervated by the same branch of the IIIrd nerve. A total IIIrd nerve palsy is usually accompanied by complete ptosis, but in these cases the presenting symptom is unlikely to be diplopia. Bilateral partial ptosis, especially in the evening, can be due to fatigue associated with myasthenia gravis. Alternatively, lid retraction, along with proptosis can be indicative of dysthyroid eye disease. Unusual patterns of lid movement can also occur in relation to Duane's syndrome, aberrant regeneration of the IIIrd nerve and dorsal mid-brain syndrome (Parinaud's).

**Prescription**

Hypermetropia may indicate a long-standing accommodative squint, whereas a prismatic correction is a sure sign of previous therapy for a binocular vision problem.

**3. Monocular Tests**

Monocular diplopia should always be excluded before attempting to further classify the diplopia.

■ **Monocular occlusion**

Monocular diplopia may be present in each eye (eg cortical diplopia) or in one eye only.

■ **VA with and without pin-hole**

Refractive monocular diplopia will normally disappear through a pinhole.

■ **Amsler chart**

Metamorphopsia could indicate an association with macula pathology.

**4. Cover test**

The cover test should be performed, at least initially, using an accommodative target for fixation. Any variation in the horizontal angle of deviation with accommodative effort needs to be taken into consideration when performing ocular motility. Binocular diplopia is normally due to misalignment of the visual axes, so a movement of the deviating eye to take up fixation will be seen on occlusion of the fixating eye. Long-standing deviations can have intermittent suppression, so the cover test could reveal a misalignment of the visual axes even when there is no subjective report of diplopia. Equally, subjects with long standing deviations can have large fusional reserves, in which case observations should be made on the direction and speed of recovery. Fixation may alternate in recent onset deviations, observation of the movement as the eyes swap fixation can be sufficient to determine the direction of the deviation without the use of a cover, or the patient can be asked to fixate one image then the other. The direction of eye movement and subjective descriptions of the images can be used to determine whether the diplopia is horizontal (crossed or uncrossed), vertical or torsional.

When the patient's symptoms suggest that the presenting diplopia is related to close work, the near cover test should be performed with the fixation target in the primary position, and with the fixation target slightly below the horizontal in a more natural reading position. This will differentiate between a defect of the convergence/accommodation system and one in which the symptoms are evoked by down gaze. 'A's and 'V's and SO palsies may fall into this latter group.

Where there is an incomitancy, the angle of deviation normally differs depending on the fixating eye, with the secondary deviation being larger than the primary. The primary deviation is the angle when fixating with the unaffected eye and the secondary deviation is the angle when fixating with the affected eye.

The cover test should be repeated with and without any AHP. Normally, the angle of deviation will reduce with the head posture and a previously manifest deviation will become latent. It is necessary to determine

whether the diplopia and manifest deviation are constant or intermittent, and whether the angle of deviation varies with fixation distance.

Various techniques have been described which use the cover test to isolate the defective muscle in an infranuclear palsy, one of which was described by Scobee in 1952 (cited by Mallett 1988).

**Scobee technique**

**I. Is the vertical deviation right hypertropia (R/L) or left hypertropia (L/R)?**

*Answer:*

R/L, the defective muscle must be a right depressor or left elevator; Right superior oblique (RSO), Right inferior rectus (RIR) or Left inferior oblique (LIO), Left superior rectus (LSR)

**II. Is the deviation greater for near or distance fixation?**

*Answer:*

near, the defective muscle must be more active on near fixation; RSO or LIO

**III. Deviation greater fixing right eye or left eye?**

*Answer:*

Fixating with the right eye (FRE) Defective muscle the RSO

A red filter in front of one eye can help determine which image is seen by which eye.

**5. Ocular motility**

This is normally used to assess binocular versional eye movements, but where there appear to be restrictions it can also be used to examine monocular ductions.

Ocular motility is generally undertaken without spectacles while the patient fixates a spot of light. There is, however, a valid argument for examining eye movements while wearing spectacles and using an accommodative fixation target in those patients who have an accommodative type deviation. Removal of spectacles makes it easier to see subtle changes and allows full ocular excursions, which may be necessary in identifying small under or over-actions. Patients with a tendency to suppress are more likely to recognise diplopia when fixating a spot light, and observation of corneal reflections ensures that neither eye is occluded by facial contours. Large angles of deviation can be observed as an asymmetry in corneal reflections, but allowances should be made for the reflexes naturally changing position during excursions due to parallax. A small shift in the position of a corneal reflex indicates a large change in eye position (a 1 mm shift in corneal reflex is equivalent to an ocular misalignment of approximately 7° or 14Δ (von Noorden, 1990)).

The alternate cover test should be performed in the different directions of gaze to reveal the full angle of the deviation. Subjective responses reporting diplopia may not be reliable in long-standing deviations with suppression. Equally, if fusion is maintained throughout the ocular

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excursions, an incomitancy may be missed.

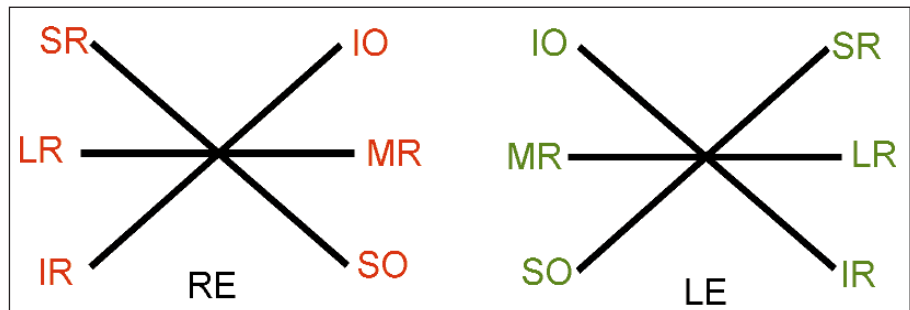
Subjective responses can be improved by the wearing of red and green goggles. Suppression is less likely and the patient can report on the direction and separation of the images. The use of a vertical bar-light as a fixation target allows for the assessment of torsion, and can be plotted as a 'diplopia chart'.

### Mechanical or myogenic versus neurogenic:

Describing an incomitancy in terms of a single muscle paresis should be done with caution, as it could lead to mis-diagnosis of the defective muscle. For example, a mechanical restriction of the SO such as Brown's syndrome, has a similar eye movement pattern to an IO paresis. Observation of the smoothness of eye movements can help differentiate between a mechanical and neurogenic defect. In a neurogenic lesion, as the eyes move into the direction of the defect, the under-acting eye will move smoothly but more slowly than the normal eye. In a mechanical deviation, the eye movements will be smooth and symmetrical until meeting the obstruction, at which point there will be an abrupt slowing of the defective eye. Another clue to differentiating a neurogenic from a mechanical lesion is by observing the diplopia. If the distal image changes in opposite directions of gaze (e.g. the image seen by the right eye is inferior on down gaze and superior on up gaze), then this indicates a mechanical deviation, and is described as a crossing of diplopia. A neurogenic lesion generally results in an under-action of the eye movement, but the eye can normally make a full excursion, if encouraged. In a mechanical restriction, the eye is often tethered, such that it cannot move into the stipulated direction. This mechanical tethering can put pressure on the globe, causing an increase in intra-ocular pressure (IOP) on an attempted movement in the direction of the restriction. A further technique used by ophthalmologists for the differentiation of a neurogenic from a mechanical palsy is the forced duction test (under anaesthetic). The globe is held with forceps and manually rotated into the direction of the under-action. If, under such conditions, the eye moves smoothly, it suggests that the under-action is neurogenic, if the movement remains restricted, it indicates a mechanical limitation. The final definitive differentiation uses electromyography. In a mechanical defect, the electromyographic activity will be normal or increased on attempting to look in the direction of the restriction, whereas the activity will be reduced with a neurogenic lesion.

If ocular motility reveals an apparent under-action of an IO, then the possibility of it being a congenital Brown's syndrome should be considered. If the eye movement pattern looks like a LR paresis, the palpebral

### Differential diagnosis of neurogenic infranuclear palsies.



**Figure 1**

The familiar plot shown in **Figure 1** shows the direction of eye movement in which specific eye muscles are most active. If any one muscle was paralysed, the direction indicated would be that of the greatest under-action. In reality, muscles supplied by the IIIrd cranial nerve are rarely affected in isolation. The SO and IO act as an antagonistic pair, elevating and depressing the eye when it is in adduction. If one muscle should become palsied (eg SO), the other would act without opposition, hence there would be over-action of the direct antagonist (eg IO). If the palsied eye was used for fixation, there would be an effect on the unaffected eye. The IR and SR work as an antagonistic pair when the eye is in abduction. In the above example, the IR of the contralateral eye would work as the synergist to the palsied muscle and hence, when fixating with the affected eye, would over-act. The SR, as its antagonist, would correspondingly under-act. The spread of this 'muscle sequelae' varies enormously between individuals dependent on a number of factors, not least the eye habitually used for fixation and the duration of the condition. The full sequelae for each muscle of each eye is shown in **Table 2**. below.

RIGHT EYE	Greatest under-action Defective muscle	Greatest over-action Contralateral synergist	Secondary over-action Direct antagonist	Secondary under-action Contralateral antagonist
Lateral Rectus	RE in Dextro-version (RLR)	LE in Dextro-version (LMR)	RE in Laevo-version (RMR)	LE in Laevo-version (LLR)
Medial Rectus	RE in Laevo-version (RMR)	LE in Laevo-version (LLR)	RE in Dextro-version (RLR)	LE in Dextro-version (LMR)
Superior Rectus	RE in Dextro-elevation (RSR)	LE in Dextro-elevation (LIO)	RE in Dextro-depression (RIR)	LE in Dextro-depression (LSO)
Inferior Rectus	RE in Dextro-depression (RIR)	LE in Dextro-depression (RSO)	RE in Dextro-elevation (RSR)	LE in Dextro-elevation (LIO)
Superior Oblique	RE in Laevo-depression (RSO)	LE in Laevo-depression (RIR)	RE in Laevo-elevation (RIO)	LE in Laevo-elevation (LSR)
Inferior Oblique	RE in Laevo-elevation (RIO)	LE in Laevo-elevation (LSR)	RE in Laevo-depression (RSO)	LE in Laevo-depression (RIR)
LEFT EYE	Greatest under-action Defective muscle	Greatest over-action Contralateral synergist	Secondary over-action Direct antagonist	Secondary under-action Contralateral antagonist
Lateral Rectus	LE in Laevo-version (LLR)	RE in Laevo-version (RMR)	LE in Dextro-version (LMR)	RE in Dextro-version (RLR)
Medial Rectus	LE in Dextro-version (LMR)	RE in Dextro-version (RLR)	LE in Laevo-version (LLR)	RE in Laevo-version (RMR)
Superior Rectus	LE in Laevo-elevation (LSR)	RE in Laevo-elevation (RIO)	LE in Laevo-depression (RIR)	RE in Laevo-depression (RSO)
Inferior Rectus	LE in Laevo-depression (LIR)	RE in Laevo-depression (RSO)	LE in Laevo-elevation (LSR)	RE in Laevo-elevation (RIO)
Superior Oblique	LE in Dextro-depression (LSO)	RE in Dextro-depression (RIR)	LE in Dextro-elevation (LIO)	RE in Dextro-elevation (RSR)
Inferior Oblique	LE in Dextro-elevation (LIO)	RE in Dextro-elevation (RSR)	LE in Dextro-depression (LSO)	RE in Dextro-depression (RIR)

fissure should be observed closely for signs of the characteristic widening and narrowing associated with Duane's syndrome.

### 6. Near point of convergence

Measuring the near point of convergence is of clear value in patients complaining of asthenopic symptoms. Assessment of convergence can also provide useful information in differentiating a supranuclear from an infranuclear defect. The anatomical centres for the generation of horizontal eye movements and for convergence are located in different areas of the brain stem. For example in a supranuclear defect of horizontal gaze adduction will be affected on versions that will be spared in convergence.

### 7. Fixation disparity

Assessment of fixation disparity aids the diagnosis of a decompensating heterophoria. Where a patient complains of intermittent diplopia, but on examination there is no manifest deviation, the presence of an uncompensated element may suggest a tendency for decompensation under conditions of visual stress. The displacement of the nonius strips can indicate which elements of the deviation are problematic, the vertical, horizontal or torsional. Beyond this, assessment of the fixation disparity is of greatest value when treating rather than diagnosing the condition.

### 8. Measurement of the angle of deviation

Measurement of the deviation in different directions of gaze may help confirm the direction of maximum misalignment of the visual axes. Different practitioners favour different methods of measurement, but it can be useful to use an accommodative target for fixation when assessing A's and V's. The horizontal angle of the deviation will vary with accommodative effort in the primary position, up and down-gaze.

In true incomitancies, the angle of deviation differs depending on which eye is fixating. In order to use the prism cover test to measure the angle when fixating with the right eye, for example, the strength of prism should be adjusted until the movement of the left eye is nulled. When fixating with the right eye, the angle of the left eye is being measured, and when fixating with the left eye, the angle of the right eye is being measured. As the secondary angle of a deviation is the greater, measurement of the angle of deviation fixating each eye in turn can help determine the eye with the defect

### Maddox double rod test

Conventional methods of measuring the angle of deviation (such as the prism cover test) can be used to measure the vertical and horizontal elements of a deviation, but cannot measure the torsional element. Maddox rods at the same orientation in front of each eye

(normally vertically orientated to produce a horizontal streak) can be used to assess the angle of torsion. If the streaks seen by each eye are not parallel, then the rods can be rotated until the streaks become parallel and horizontal, thus giving a measure of the rotation required and hence the torsion. This test is maximally dissociating and can produce erroneous results (possibly as a result of small angles of head tilt). Bagolini lenses can be used in the same way, but this method tends to underestimate the angle of torsion, due to fusion. In a IVth nerve palsy, torsion greater than 10° is often thought to indicate a bilateral rather than unilateral condition, but the validity of this general rule has been questioned (von Noorden, 1990).

### 9. Bielschowsky

Due to the development of the muscle sequelae, the eye movement pattern in a longstanding SO palsy in one eye can be difficult to differentiate from a SR palsy of the other eye. Bielschowsky devised the head tilt test in 1900. A positive Bielschowsky test can confirm the culprit as the SO, but a negative result is inconclusive. Normally, as the head is tilted towards the right shoulder, for example, the right SR and right SO work in partnership to intort the eye, the opposing vertical actions of the two muscles cancelling out. In lower animals, this action re-orientates the eye to the vertical but, in man, the system is less efficient. If a patient has a SO palsy, as the head is tilted towards the affected side, the SR acts unopposed, so it not only intorts the eye but also elevates it.

To perform the test, seat the patient upright, maintaining steady fixation straight ahead at a distance of 3m, so that fixation doesn't favour either the SO or SR. Tilt the head towards the eye with the suspected SO palsy (the hypertropic eye) and if the vertical angle of the deviation increases, as shown for the RSO in **Figure 2**, the defective muscle is confirmed as the SO (von Noorden 1990).

In 1958, Parks used this information to devise a three step test for differentiating the four vertically acting extra-ocular muscles.

**Figure 2** The Bielschowsky head tilt test can be used to differentiate a RSO and palsy from an LSR palsy. When the head is straight, the affected right eye is slightly hypertropic, esotropic and extorted. On tilting the head towards the right shoulder (the affected side), the angle of the right hypertropia increases. This would not occur in a Left Superior Rectus Palsy. On tilting towards the left shoulder, there is little difference in the vertical angle of deviation.



### I. Is the deviation R/L or L/R in the primary position?

*Answer:*

R/L, muscles involved include R depressors and L elevators:  
RIR, RSO or LSR, LIO

### II. Does the vertical deviation increase on laevo or dextro version?

*Answer:*

laevo version, muscles involved active on looking to the left:  
RSO, LSR

### III. Does the vertical deviation increase on head tilt to the left shoulder or the right?

*Answer:*

to the right:  
defective muscle RSO.

### 10. Fusional Reserves

Measurement of fusional reserves can be of diagnostic value when differentiating a long standing vertical muscle palsy from one of recent onset. Congenital SO palsies, for example, can have vertical fusional reserves in excess of 10°, whereas a recent onset deviation will usually have a normal vertical fusion range (4° - 6°). Vertical fusion ranges can also increase over a long period of gradual change in the direction of the visual axes, such as in dysthyroid eye disease.

Measurement of the vertical fusion range can be done with the patient fixating either a near or a distant target. Patients with long-standing deviations may not appreciate diplopia after fusion breaks, so it may be useful to introduce visual controls to ensure neither eye is being suppressed. If a pen torch is used to illuminate a near fixation target, and obliquely orientated Bagolini lenses are positioned in front of each eye, the streaks produced by the Bagolini lenses can act as a control.

### 11. Past Pointing

Past pointing can be used to differentiate a recent onset from a long-standing condition. On occlusion of the unaffected eye, the patient is asked to rapidly look at, and point towards, an object in the field of action of the palsied muscle. In recent onset incomitancies, the input required to look at the object will be greater than normal, so the object will be perceived as more peripheral than its actual position, and the patient will tend to point towards a more eccentric location. This test is improved if the patient cannot see their hand, not allowing them to use visual feedback to correct its direction of movement. Over time, the ocular motor feedback adjusts to the increased neurogenic input, and object localisation becomes more accurate.



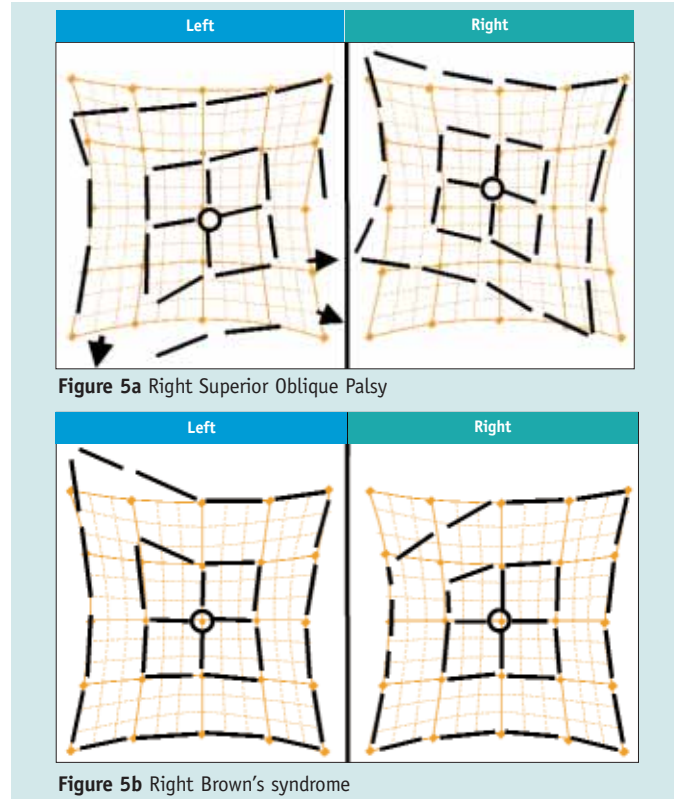
head centred on the central fixation spot. Patients with longstanding AHPs often find it difficult to sit with their head straight, so a chin rest should be used to ensure the correct head position is maintained. Ideally, plot the centre position first, then the 15° fixation points, and finally as many of the 30° points as can be seen without moving the head. On the latest computerised charts, the small squares are not necessarily the conventional 5°, and hence the inner and outer plots are equally re-scaled. Ensure the plot is marked with the appropriate scaling.

There are a number of basic rules for interpreting a Hess plot:

1. The smaller field belongs to the eye with the defect.
2. Neurogenic pareses will show the muscle sequelae to a greater or lesser extent (dependent on the duration

of the condition and the eye used for fixation). This is illustrated in the Hess plot for a RSO paresis shown in **Figure 5a**. The largest under-action is normally in the direction of action of the paretic muscle and the largest over-action is normally the contralateral synergist.

3. Mechanical defects show a compressed field. There is not normally an obvious over-action of the direct antagonist, nor under-action of the contralateral antagonist, so the effects of the defect are limited to the direction of action of the mechanical restriction. This is illustrated in the Hess plot for Brown's syndrome shown in **Figure 5b**. The most obvious feature of a mechanical defect is normally the marked over-action of the contralateral synergist.



## Module 2 Part 10

Having discussed an appropriate diagnostic routine for patients presenting with diplopia, the following is a summary of causative factors and their differential characteristics.

### MONOCULAR DIPLOPIA:

- **Diagnostic Feature**  
Diplopia remains on occlusion of one eye.

### REFRACTIVE:

Ocular media, lenticular (Brown, 1993) or corneal (Campbell, 1998). Astigmatic or ametropic (Coffeen and Guyton, 1988). Lid position (Ford et al., 1997), spectacle induced (bifocal segment), contact lens induced (Morris, 1991).

- **Diagnostic Feature**  
Diplopia disappears through a pinhole.

### RETINAL:

Maculopathy or retinal irregularity (Lepore and Yarian, 1986).

- **Diagnostic Feature**  
Normally associated with metamorphopsia

### CORTICAL DIPLOPIA:

Very rare. Can be reported as diplopia (Kasman and Ruprecht, 1995) or polyopia (Cornblath et al., 1998).

- **Diagnostic Features**  
Generally associated with other cortical dysfunction. Diplopia or polyopia remains on occlusion of each eye, and is not resolved by viewing through a pinhole.

### STRABISMIC:

Rare. Occurs after squint surgery in subjects with well-established Abnormal Retinal Correspondence (causing binocular triplopia) or after treatment for amblyopia (von Noorden, 1990).

- **Diagnostic Feature**  
Associated with long standing concomitant deviation and orthoptic therapy. Normally there is no confusion between the 'real' and 'diplopic' image.

### PSYCHOGENIC:

(Records, 1980).

- **Diagnostic Features**  
Symptoms inconsistent with any other diagnosis. Separation of the images does not change with viewing distance.

### BINOCULAR DIPLOPIA

- **Diagnostic Feature**  
Diplopia resolved by occlusion of either eye.

### PHYSIOLOGICAL:

Patients suddenly become aware of physiological diplopia of objects in front of or behind fixation.

- **Diagnostic Feature**  
Diplopia normally noticed fleetingly and only when tired.

### REFRACTIVE:

Edge effects of high prescription spectacles or bifocals. Prismatic effect of ill-fitting spectacles. Contact lens edge effects.

- **Diagnostic Features**  
Consistent image separation in different directions of gaze. Disappears on removal of refractive correction.

### RETINAL:

Maculopathies, resultant aniseikonia can make fusion difficult (Benegas et al., 1999).

- **Diagnostic Features**  
Retinal signs, metamorphopsia, aniseikonia.

### CONCOMITANT:

#### CONVERGENCE:

Convergence insufficiency.

- **Diagnostic Features**  
Little or no heterophoria on near or distance fixation. Poor binocular convergence.

Convergence/accommodative spasm can be indicative of a neurological pathology (Raymond and Crompton 1990).

- **Diagnostic Features**  
Increased esotropia on distance fixation. Miosis when both eyes are open. Apparent Myopia.

#### DECOMPENSATING HETEROPHORIA:

- **Diagnostic Features**  
Diplopia normally associated with other asthenopic symptoms (e.g. headache) and tends to occur following a specific visual activity. Poor fusional reserves and measurable 'slip' on assessment of fixation disparity.

#### HETEROTROPIA:

Diplopia is rarely a presenting symptom in children. A recent onset concomitant deviation is most frequently the result of uncorrected hypermetropia or an imbalance in the accommodative convergence relationship, but other factors may be involved. Concomitant strabismus can be the presenting symptom for retinoblastomas (Abramson et al., 1998) or intracranial tumours (Williams and Hoyt, 1989).

- **Diagnostic Features**  
Difficult to differentiate the benign from the sinister, so all children with a concomitant squint of unknown aetiology should have a thorough fundus examination and be treated with caution.

### LONG-STANDING STRABISMUS:

Constant, irreversible diplopia can result from over-zealous occlusion therapy in the older child or anti-suppression exercises in subjects with poor binocular function.

- **Diagnostic Feature**  
History of previous orthoptic treatment.

### As AND Vs:

The 'alphabet patterns'.

- **Diagnostic Feature**  
Difference in horizontal angle of deviation greater than 10° between up and down gaze.

### INCOMITANT:

Angle of deviation differs in different directions of gaze and fixating either eye.

### LONG-STANDING:

Symptoms caused by long-standing incomitancies can be distressing, but are not an emergency. The possibility of a recent onset lesion occurring in addition to a longstanding deviation cannot be excluded.

- **Diagnostic Features**  
Vague description of symptoms and their onset. Patients are often unaware of a long-standing AHP and may have associated facial asymmetry. Can have increased vertical motor fusion range.

### RECENT ONSET:

Establish a cause if possible, and refer with a degree of urgency depending on the diagnosis.

- **Diagnostic Feature**  
Very specific description of symptoms and their onset. Symptoms very distressing. Any AHP and may cause a neck ache.

### Incomitancies due to neurological disease;

#### SUPRANUCLEAR:

Neurological lesions above the brain stem nuclei may cause isolated defects of conjugate eye movements (resulting in a gaze palsy) or of vergences. For example, a horizontal gaze palsy could affect conjugate eye movements to the right, so there would be no adduction of the left eye on attempted versions, but convergence may be normal. Supranuclear lesions are not normally associated with diplopia, although there are exceptions, such as in skew deviation, in which there is a vertical misalignment of the visual axes and vertical diplopia (Acierno, 2000). ➔

#### SUPRANUCLEAR: (continued)

##### ■ **Diagnostic Feature**

*Eye movement dysfunction may selectively affect saccades or smooth pursuit, leaving vestibular reflexes intact. Defects are normally binocular and cannot be isolated to a specific muscle.*

#### INTERNUCLEAR:

Often associated with oscillopsia on lateral gaze, there may not be recognition of diplopia. A defect affecting the medial longitudinal fasciculus communication between the VIth nerve nucleus and the IIIrd nerve MR subnucleus. Internuclear ophthalmoplegia (INO) normally indicates MS or brain stem vascular disease (O'Boyle et al., 1992). An INO can have VIth nerve involvement (One and a half syndrome) or can be associated with XOT (Wall eyed syndrome) (Brazis and Lee 1999).

##### ■ **Diagnostic Feature**

*Weakness of one eye on adduction with nystagmus of the contralateral abducting eye.*

#### NUCLEAR:

Nuclear lesions can affect the entire nucleus or, in the case of the IIIrd nerve, isolated sub-nuclei (Harrison and Wirtschafter, 1999). In a complete, unilateral IIIrd nerve nucleus lesion, the ipsilateral MR, IR and IO are affected, the contralateral SR, bilateral ptosis and internal ophthalmoplegia (Glaser 1999).

##### ■ **Diagnostic Feature**

*Difficult to differentiate from an infranuclear lesion. In a complete IIIrd the contralateral SR is effected, and both levators.*

#### INFRANUCLEAR:

The most common neurogenic cause of binocular diplopia. There are numerous causes, mainly vascular (including diabetes and migraine), tumours (benign or malignant) or trauma. Isolated VIth and IVth nerve palsies occur the most frequently. A total IIIrd affects the ipsilateral MR, IR, IO, SR, levator and internal ocular musculature. Classically, a total IIIrd due to microvascular anomalies, such as diabetes, has pupil sparing. A partial IIIrd with pupillary involvement may be due to a compressive lesion. A posterior communicating artery aneurysm can cause a compressive lesion, normally associated with pain. If such an aneurysm bursts, the consequences can be fatal. Infranuclear palsies isolated to only one muscle supplied by the IIIrd nerve are rare, but not unheard of (von Noorden GK and Hansell, 1991). A total IIIrd can recover abnormally to

#### INFRANUCLEAR: (continued)

form a condition known as aberrant regeneration. Classically, this results in adduction on attempted up gaze, lid retraction on down gaze and miosis on attempted adduction.

##### ■ **Diagnostic Features**

*Classic picture of ocular motor paresis and resultant sequelae. Vergences, conjugate saccades and pursuit affected equally. A painful IIIrd nerve lesion should be referred as an emergency*

#### NEURO-MUSCULAR JUNCTION:

Myasthenia Gravis (Barton and Fouladvand, 2000) and other myasthenic like syndromes. Myasthenia can present with almost any single muscle affected. The deviation is variable, it may present as completely different pareses on consecutive visits. The condition improves following rest and is associated with general systemic weakness and rapid fatigue. An autoimmune disease, the diagnosis is confirmed following a rapid but short-term improvement in muscle function following an injection of Tensilon.

##### ■ **Diagnostic Features**

*Normally inconsistent with a single muscle palsy. Associated with symptoms of general fatigue. Signs of eye lid fatigue include worsening ptosis on prolonged upgaze.*

#### MYOGENIC:

Systemic conditions directly affecting the extra-ocular muscles, and producing a mechanical type restriction of movement. Dysthyroid eye disease (Grave's ophthalmopathy) (von Noorden, 1990). Orbital myositis. Ocular myopathies (including progressive external ophthalmoplegia) (Glaser, 1999).

##### ■ **Diagnostic Features**

*Other signs of orbital inflammatory or myopathic disease. Restrictions inconsistent with single muscle paresis. Sequelae may not affect ipsilateral or contralateral antagonists.*

#### CONGENITAL:

Patients with congenital ocular motor disorders rarely experience distressing diplopia. Congenital conditions can take on almost any form of nerve or gaze palsy, most commonly IVth and VIth, Moebius syndrome (combined VIth and VIIth nerve palsy) (Lammens et al., 1998) or Duane's syndrome (Duane, 1996)

##### ■ **Diagnostic Features**

*Rarely symptomatic, although weak suppression on lateral gaze can result in symptomatic diplopia. Long-standing AHP to maintain binocular vision may result in facial asymmetry. Congenital IVth normally associated with extended vertical fusion range.*

#### Incomitancies due to mechanical restrictions:

##### ACQUIRED:

Acquired Brown's syndrome (Saunders et al., 1990).  
Blow-out fracture (Al-Qurainy et al., 1991).  
Orbital space occupying lesion (eg cellulitis, tumour) (Vernet et al., 1993).

##### ■ **Diagnostic Features**

*Similar to myogenic palsies. Sequelae do not affect ipsilateral or contralateral antagonists and can get 'crossing of diplopia'. Other signs of orbital involvement, eg hyperaemia, exophthalmos, enophthalmos.*

##### CONGENITAL:

Brown's syndrome (Brown, 1973).  
Johnson's adherence syndromes (von Noorden, 1990). The tendon sheaths of the IO and LR or of the SO and SR become adherent.

##### ■ **Diagnostic Features**

*Rarely symptomatic. Diplopia normally avoided by suppression. Smooth eye movements up to the point of restriction, then an abrupt onset under-action. In Brown's syndrome, there may be full movement following an audible click.*

In summary, many patients complaining of diplopia will have conditions that can be appropriately managed in practice. For example, congenital SO palsies in their first pair of bifocals or varifocals may become symptomatic, a situation that can be relieved by using separate readers. Other cases will need routine referral, for example long-standing deviations which are cosmetically unacceptable may be assisted by surgery. Those for whom the condition is more sinister and require neuro-ophthalmologic investigation are, fortunately, uncommon. The consequences of inappropriate action, however, can be life threatening. When a patient presents complaining of diplopia, consider the following:

1. Is it monocular or binocular?
2. Is it refractive or due to misalignment of the visual axes?
3. Is it an imbalance of the accommodative/convergence mechanism or the ocular motor system?
4. Is it long-standing or of recent onset?
5. Was the onset gradual or acute?
6. Is there any associated cause, pain systemic symptoms or pathology?

In an adult or a child, recent, acute onset diplopia due to a concomitant or incomitant deviation with an uncertain aetiology should be referred urgently for a neuro-ophthalmologic investigation.

## Module 2 Part 10

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### About the Author

Dr Alison Finlay worked for several years as an orthoptist before studying optometry. After a few years in practice, she moved into research at Imperial College, London. Following completion of a PhD, she took up a lectureship at City University.

## Multiple choice questions – Please note there is only **ONE** correct answer

### The differential diagnosis of diplopia

1. **In a recent onset incomitancy due to a neurogenic lesion, the diplopia is usually described as:**
  - a. 'ghosting' rather than a true diplopia
  - b. gradual in onset
  - c. worse in specific directions of gaze
  - d. present fleetingly when tired
2. **Concomitant esotropia presenting in childhood**
  - a. never presents with diplopia
  - b. can be caused by neurological pathology
  - c. is normally associated with myopia
  - d. is always treated effectively by refractive correction
3. **Which of the following is not normally associated with an abnormality of eye lid movement?**
  - a. Dysthyroid eye disease
  - b. Duane's retraction syndrome
  - c. Aberrant regeneration of the IIIrd nerve.
  - d. Multiple Sclerosis
4. **What is the most likely diagnosis of a patient presenting with an IR paresis and a history of general fatigue?**
  - a. Myasthenia Gravis
  - b. Moebius syndrome
  - c. Progressive External Ophthalmoplegia
  - d. Diabetes Mellitus
5. **A patient with a long standing incomitancy and an abnormal head posture will normally**
  - a. move the eyes into the direction of action of the defective muscle
  - b. avoid diplopia by suppression, both with and without the head posture
  - c. constitute an emergency
  - d. have larger than normal amplitudes of motor fusion
6. **Which of the following is a feature of a neurogenic but NOT a mechanical ocular motor defect?**
  - a. Crossing of diplopia between up and down gaze
  - b. Limitation of eye movement on forced ductions
  - c. Over-action of the contralateral synergist
  - d. Over-action of the direct antagonist
7. **The clinical picture of Brown's syndrome can be confused with:**
  - a. an Inferior Oblique palsy
  - b. a Superior Oblique palsy
  - c. Duane's retraction syndrome
  - d. a blow-out fracture
8. **A 1 mm shift in corneal reflex is equivalent to an ocular misalignment of approximately:**
  - a. 4<sup>Δ</sup>
  - b. 14<sup>Δ</sup>
  - c. 7<sup>Δ</sup>
  - d. 2<sup>Δ</sup>
9. **Beilschowsky head tilt test confirms the left superior oblique as the defective muscle when ...**
  - a. right hypertropia increases on head tilt to the right.
  - b. left hypertropia increases on head tilt to the right.
  - c. left hypertropia increases on head tilt to the left.
  - d. right hypertropia increases on head tilt to the left
10. **When interpreting a Hess chart:**
  - a. distal point always belongs to the affected eye.
  - b. the smaller field is the field of the affected eye
  - c. the larger field is the field of the affected eye
  - d. the greatest over action is the contralateral antagonist
11. **Supranuclear lesions generally:**
  - a. present with distressing diplopia
  - b. spare the vestibulo-ocular reflexes
  - c. affect a single ocular muscle
  - d. cause a painful red eye
12. **A painful, partial IIIrd nerve palsy should be treated as an emergency, because in the worst case scenario, this could lead to:**
  - a. permanent diplopia
  - b. loss of vision in the affected eye
  - c. increased pain
  - d. death

An answer return form is included in this issue.  
It should be completed and returned to:  
CPD Initiatives (NOE10),  
OT, Victoria House, 178-180 Fleet Road, Fleet,  
Hampshire, GU13 8DA by November 1.