Introduction

Community optometrists should be able to recognise when an incomitant strabismus is of recent onset and requires urgent referral for ophthalmological and neurological investigation. They must also be able to investigate and diagnose when an incomitant deviation is longstanding to avoid unnecessary referral when treatment is not required. Furthermore, it is important for community optometrists to be able to detect incomitant deviations in childhood and manage appropriately.

This two-part series reviews the causes and management of incomitant strabismus that are likely to present to community optometry. This first article introduces the characteristics of incomitant strabismus and the investigative procedures that aid differential diagnosis. Diagnosis and management of neurogenic palsies is also included. The second article will cover the diagnosis and management of mechanical, myogenic and letter pattern deviations.

What is incomitant strabismus?

An incomitant strabismus is one in which the angle of deviation changes in different positions of gaze; this differs to a concomitant strabismus which remains the same size in all directions of gaze (e.g., accommodative esotropia, intermittent exotropia). Incomitant strabismus is classified as: neurogenic (relating to a problem of the nervous supply to the ocular muscle), myogenic (a weakness of the ocular muscle itself) and mechanical (a physical limitation of the ocular muscle) according to the underlying cause. Incomitant deviations can also occur in the form of letter pattern deviations typically due to anatomical differences in the muscle insertions or orbital structure. It is important to be able to differentially diagnose between incomitant and concomitant strabismus as a newly acquired incomitant strabismus may signal a condition that can affect the patient’s general health and have a potentially life-threatening aetiology. Incomitant strabismus occurs where there is a limitation of ocular movement that typically causes the angle of deviation to increase as the eyes are turned in the direction of the limitation, and decrease as the eyes are turned in the opposite direction. There are some exceptions to this rule when the cause is mechanical and the movement can be limited in the opposing direction of the limitation.

The degree of the limitation can also be classified as paralysis, where no movement of the eye in a particular direction is possible, or paresis, where there is some, limited movement of the eye in a particular direction. However, the term generally accepted to describe any limitation of ocular movement, whether paralysis or paresis, is palsy. Incomitant strabismus can also be classified by the
age of onset, e.g. whether they are congenital or acquired. Congenital palsies are usually due to a developmental anomaly of the ocular motor system, such as the anatomy of the muscle itself, or a defect in the nervous system supplying the muscle. Acquired palsies are caused by an injury or disease of the ocular motor system that occurs after the first six months of life.

Characteristics of incomitant strabismus

Both congenital and acquired incomitant deviations may be fully or partially controlled by a compensatory head posture and/or the patient’s fusional reserves which results in the development and maintenance of binocular single vision. The principle of a compensatory head posture is to place the eyes in the position of least deviation to develop and maintain binocular single vision, avoiding looking in an area of pain or diplopia, and to centralise the field of binocular single vision. Less frequently, a compensatory head posture may occur where the eyes are placed in the position of greatest deviation, to have the greatest separation of images, with the aim of ignoring the image furthest away. Compensatory head postures consist of face turns, head tilts, and chin elevations or depressions, with some conditions encompassing all three. The presence of a compensatory head posture should be noted and should trigger further investigation for the presence of an incomitant strabismus.

Some incomitant deviations may not be compensated and will give rise to a manifest strabismus in primary position, as well as increasing in size in the direction of the affected muscle. They may also be accompanied with symptoms such as diplopia and/or pain if it is of recent onset in adulthood, or they can be symptom-free in children (or in adulthood if onset was in childhood) with the presence of suppression.

Another characteristic of incomitant deviations is the presence of muscle sequelae. These are a result of the initial limitation of movement affecting one muscle, followed by a series of muscle overactions and underactions/inhibitions according to the laws that govern ocular movement (Hering’s and Sherrington’s Laws) (detailed in Tables 1A and B). If the condition is of recent onset, only the first two muscle sequelae will be present, which will make diagnosis of the affected muscle more definite. Muscle sequelae will be established if the palsy has been present for a longer duration and can often make diagnosis of the initial muscle palsy difficult.

<table>
<thead>
<tr>
<th>Muscle sequelae order</th>
<th>Ocular movement law</th>
<th>When present?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary underaction/muscle palsy</td>
<td>Hering’s Law (equal innervation)</td>
<td>From the onset</td>
</tr>
<tr>
<td>Overaction of contralateral synergist</td>
<td>Sherrington’s Law (reciprocal innervation)</td>
<td>After a period of time (usually a few months)</td>
</tr>
<tr>
<td>Overaction (contracture) of ipsilateral antagonist</td>
<td>Hering’s and Sherrington’s Law</td>
<td>After a period of time (usually a few months)</td>
</tr>
</tbody>
</table>

Table 1A: Muscle sequelae development following a primary muscle palsy. Contralateral = opposite side; ipsilateral = same side; synergist = yoke muscle; antagonist = opposing muscle

Investigation of incomitant strabismus

A comprehensive eye examination should be carried out to establish the type of incomitant strabismus present, to differentially diagnose neurogenic from myogenic and mechanical palsies, and to determine the type and urgency of management required. The investigative procedures should begin with a careful case history covering all ocular symptoms and any changes in general health. Poor health can cause decompensation of a congenital incomitant strabismus and some health conditions can cause incomitancy, e.g. thyroid disorders, vascular conditions (diabetes/high blood pressure/high cholesterol), myasthenia gravis and recent trauma. Diplopia is often a symptom reported with incomitant strabismus and should be given thorough consideration. Marked diplopia of sudden onset, which is often distressing and is often accompanied by pain around the eye, is a sign of a recently acquired third nerve palsy that can have life-threatening neurological consequences and must be referred immediately for medical investigation.

It should be confirmed whether the reported diplopia is monocular or binocular by asking the patient to close each eye in turn. Monocular diplopia occurs as a result of media changes, typically cataracts or uncorrected astigmatism, and is not related to incomitant strabismus or a binocular vision problem. The patient should also be asked whether the diplopia is horizontal or vertical, or, if it is a combination of the two, which has the greater element? Horizontal diplopia is typically associated with a limitation of the horizontally acting muscles (lateral and medial recti), while vertical diplopia is typically associated with a limitation of the vertically acting muscles (superior and inferior obliques, superior and inferior recti). Torsional diplopia (i.e. where there is image tilt) is typically associated with the oblique muscles. If the patient describes the diplopia getting worse with a certain direction of gaze, this is indicative of an incomitant strabismus and can be further investigated using ocular motility, particularly in the position described, to identify the affected muscle; this is described later in the article. Diplopia that gets worse during near vision can be associated with a weakness of the superior oblique muscle which acts when looking down and in (often referred to as the reading muscle). By contrast, diplopia worsening on distance gaze is more likely to be the lateral rectus muscle as a result of a weakness of abducting/diverging when attempting to fixate in the distance. Some patients may complain that their diplopia varies depending on the time of day e.g. in myasthenia gravis and thyroid eye disease; these will be discussed in Part 2.

Carrying out the cover test in primary position can expose an incomitancy as the size of the deviation will alter depending on which eye is fixating. This is due to the extra innervation required of the paretic muscle to fixate when the fellow eye is covered. The non-paretic eye will then overact (behind the cover) due to the extra innervation (Hering’s Law) and will have a greater compensatory
movement once the cover is removed. Other tests of binocular vision to assess the duration of the condition include an assessment of fusional reserves and suppression. A longstanding incomitancy can have abnormally large fusional reserves, particularly vertically in the case of a longstanding fourth nerve. Suppression is more likely to be present in a longstanding congenital deviation, although it may be present in a recently acquired palsy in a young child. A thorough ocular examination should be carried out to rule out any related pathology. It is also important to assess the pupillary reflexes and accommodation due to the involvement of the sphincter pupillae and ciliary muscles with the third nerve. A gross observation of the patient should be carried out while taking a case history, looking for any signs of ptosis, proptosis, obvious ocular injury and difficulties with walking or speech that may help with diagnosis.

Causes of incomitant strabismus

There are many causes of incomitant strabismus that could be encountered by community optometrists. This article will cover the neurogenic causes; Part 2 of this series will cover mechanical and myogenic conditions and letter pattern deviations that optometrists may come across in community practice.

Neurogenic palsies

The third, fourth and sixth cranial nerves are responsible for the control of ocular movement. The third nerve is known as the oculomotor nerve and supplies the majority of the extraocular muscles: the superior, inferior and medial recti, and the inferior oblique. The third nerve also controls the sphincter pupillae for pupillary constriction, the ciliary muscle for accommodation and the levator palpebrae superioris for lifting the upper eyelid. The fourth nerve is known as the trochlear nerve and supplies only one muscle, the superior oblique. The sixth nerve, known as the abducens nerve, also supplies only one muscle, the lateral rectus (the abducting muscle).

Testing the action of the muscles

Figure 1 depicts the primary direction of eye movement required to test the function of the six extraocular muscles. The lateral rectus is the abducting muscle (away from the nose) and the medial rectus is the adducting muscle (towards the nose) so their function is tested in abduction and adduction respectively. Testing of the elevators and depressors requires them to be placed in either abduction or adduction before elevation or depression. This isolates the muscles so that in these positions they are the primary muscle acting to move the eye. To test the actions of the superior and inferior recti, the eye must be abducted prior to elevating and depressing respectively. Similarly, to test the superior and inferior obliques, the eye must be adducted before depressing and elevating respectively. Knowing the actions of the muscles and recording where there is limitation of movement will help to identify the affected nerve.

Third nerve palsy

A third nerve palsy can be complete, affecting all the muscles supplied by this nerve, or partial, affecting only some of the muscles due to the superior and inferior branching of the nerve. A complete acquired third nerve palsy will present with pain around the eye of sudden onset, headache, ptosis, a dilated pupil and diplopia (if the ptosis is not fully covering the pupil). The eye will be divergent due to the spared lateral rectus muscle, and depressed due to the spared superior oblique muscle, commonly referred to as the “down and out” position (Figure 2). A partial third nerve palsy will have some, but not all, of the symptoms and the eye position will depend on which muscles have been affected/spared. If there is pupil involvement, an aneurysm of the posterior communicating artery should be suspected; this is commonly referred to as a ‘medical third’. Regardless of whether the pupil is involved or not, if a third nerve palsy is suspected by the community optometrist, the patient requires an emergency referral to hospital to rule out a life-threatening aneurysm.

Figure 2: ‘Down and out’ eye position associated with 3rd nerve palsy and accompanying ptosis. Image adapted from http://www.mircophth.com/ptosis/thirdnervepalsy.html

Fang et al. reported that the most common causes of third nerve palsy presenting to their hospital in the United States were microvascular (e.g. diabetes/hypertension: 42%), trauma (12%), compression from tumour (10%) and compressive aneurysm (6%).

Pupil involvement was rarely reported in the cases of microvascular cause (16%) but was reported in the majority of traumatic (71%) and compressive cases (64%). They also reported that the incidence of an acquired third nerve palsy was seven times higher in individuals over 60 years of age when compared with those under 60 years.

After referral to the hospital, the patient will undergo neurological investigation to determine the underlying cause of their third nerve palsy and appropriate treatment will be put in place. If the cause is microvascular, recovery of the function of the third nerve is highly likely after the underlying condition has been treated (95%).

However, if the cause is traumatic, from a compressive tumour or as a result of aneurysm, recovery of the third nerve function is much less likely (19–36%).

If the patient reports diplopia (where ptosis is absent or incomplete), prisms are usually of little help due to the complexity of the deviation; therefore, an occlusive contact lens or patch may be issued. When the condition is stable and has not resolved with time, surgery is indicated to correct the strabismus and any associated ptosis. Surgery for third nerve palsy is complicated and is often performed for cosmetic reasons rather than for regaining any useful binocular single vision.

Congenital palsies of the third nerve are rare. They are often caused by the complete absence of the nerve and have other associated neurological deficits; as a result, they are unlikely to present to the community optometrist in the first instance.
Sixth nerve palsy

The sixth nerve supplies only the lateral rectus; therefore, a lesion affecting this nerve will cause an inhibition of abduction only. The sixth nerve originates from the back of the brain (from the pons) and, as it has a long way to reach the lateral rectus, it is vulnerable to injury. The peak incidence of sixth nerve palsy is between 60 and 70 years of age. The cause of an acquired sixth nerve palsy is dependent on the age of the patient; these are outlined in Table 2. The majority of sixth nerve palsies in older adults are as a result of vascular problems such as diabetes and hypertension, or a combination of both. Children with sixth nerve palsies are often found to have a congenital origin which may arise from birth trauma due to forceps or vacuum delivery. A congenital sixth nerve palsy needs to be differentially diagnosed from Duane’s retraction syndrome (DRS) which presents with similar signs (discussed in more detail in Part 2). Typically, DRS presents with a smaller esotropia in primary position than a sixth nerve palsy and will be accompanied by narrowing of the palpebral fissure and retraction of the globe on adduction, however, globe retraction can be hard to detect in infancy. Acquired sixth nerve palsies in children are reported to be most commonly caused by neoplasm (brain tumour), therefore, urgent referral for neuroimaging is important where sixth nerve palsy is suspected.

A patient with an acquired sixth nerve palsy will attend the optometric practice complaining of a sudden onset of horizontal diplopia which is typically worse on distance fixation compared with near (the opposite of a fourth nerve palsy). They may also present with a compensatory head posture consisting of a face turn to the affected side or report that the diplopia is relieved if they turn their head to a particular side. Recent onset nerve palsies typically present with an awkward/uncomfortable head posture, compared with those where the patient is not overtly aware in longstanding conditions. An esotropia will be present in primary position and ocular motility will show marked reduction of abduction of the affected eye (Figure 3). Esotropia is a common finding in children and sixth nerve palsy should always be considered as a possible cause.

Table 2: Common causes of acquired sixth cranial nerve palsy by age of onset

<table>
<thead>
<tr>
<th>Age at onset</th>
<th>Common causes</th>
</tr>
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<tbody>
<tr>
<td>Childhood</td>
<td>Neoplasm</td>
</tr>
<tr>
<td></td>
<td>Trauma</td>
</tr>
<tr>
<td></td>
<td>Raised intracranial pressure</td>
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<tr>
<td></td>
<td>Inflammatory conditions</td>
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<tr>
<td></td>
<td>Post viral infection</td>
</tr>
<tr>
<td>Young adults</td>
<td>Neoplasm</td>
</tr>
<tr>
<td></td>
<td>Multiple sclerosis</td>
</tr>
<tr>
<td></td>
<td>Diabetes</td>
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<tr>
<td></td>
<td>Post viral infections</td>
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<tr>
<td></td>
<td>Trauma</td>
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<tr>
<td>Older adults</td>
<td>Hypertension</td>
</tr>
<tr>
<td></td>
<td>Diabetes</td>
</tr>
<tr>
<td></td>
<td>Neoplasm</td>
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</tbody>
</table>

In community practice, if a sixth nerve palsy is suspected in a child, an urgent referral to orthoptics and ophthalmology (within one week) should be instigated to determine whether the condition is congenital or acquired, and to establish the underlying aetiology to allow for systemic treatment to commence. The ophthalmic goal is to try to regain binocular vision and promote normal visual development. This may require the temporary use of prisms while a period of time is given for the condition to self-resolve, if acquired. Occlusion therapy may be required to treat any amblyopia present and strabismus surgery may be indicated where a compensatory head posture is uncomfortable or where the esotropia is cosmetically unacceptable. Depending on the cause of the sixth nerve palsy, the ophthalmic issues may be of lesser importance, for example in children diagnosed with a neoplasm; however, with improving survival rates, maximising visual function is important for the child’s future development.

An urgent referral is also required for an adult with a sudden onset suspected sixth nerve palsy to investigate the underlying cause and instigate treatment. The condition will be monitored by orthoptics for a period of time to observe recovery of the nerve function. King et al. reported that almost 85% of their study group who were likely to resolve had done so within four months after treatment had commenced. Fresnel prisms may provide a helpful relief from diplopia in the interim, or patients can opt for an occlusive lens/patch. Similar to third nerve palsies, the prognosis for spontaneous recovery is better when the cause is vascular, compared with traumatic or compressive tumours.

Fourth nerve palsy

The fourth nerve controls only the superior oblique muscle and is responsible for intorting, depressing and abducting the eye. A palsy of the fourth nerve is one of the most common causes of vertical and torsional diplopia and is the most common nerve palsy to present to community practice. Due to its long pathway from the back of the brain to the eye, it is vulnerable to injury. A fourth nerve palsy can often present with a sudden onset of vertical diplopia (may also have an oblique/torsional element). This may be as a result of an acquired palsy, or as a result of decompensation of a longstanding congenital palsy (the onset just seems sudden to the patient as the diplopia was not there before). The diplopia usually worsens when reading or when looking in down-gaze (e.g. eating/going down stairs). Typically, a patient with a decompensating congenital palsy will have a comfortable compensatory head posture (usually a head tilt with chin depression) that they are unlikely to be aware of. They often have very large fusional reserves far exceeding the normal 1-2Δ range. Decompensation usually occurs with age when the deviation becomes too difficult to control; the patient may also have symptoms of asthenopia and headaches. An acquired palsy often occurs as a result of head trauma, typically to the back of the head, and can occur with relatively minor injuries. Where trauma hasn’t occurred, other vascular conditions such as diabetes and hypertension need to be investigated and, in rarer cases, neoplasm.

Carrying out a cover test will reveal a hypertropia of the affected eye in primary position. The size of the deviation will worsen and the eye will elevate if the patient’s head is tilted to the affected side. There may only be a hyperphoria present when the patient’s head is tilted to the unaffected side. This is known as the Bielschowsky head tilt test and is diagnostic of a fourth nerve palsy (Figure 4). Bilateral palsies frequently occur as a result of a head trauma and may cause a

Figure 3: Ocular posture in primary position and horizontal eye movements in sixth nerve palsy, image adapted from Curi et al.
small hypertropia in primary position when the extent of the palsy is asymmetrical. Tilting the head to either side will result in a worsening of the hyper deviation. Ocular motility will reveal underaction of the affected eye(s) in the down and in position; typically, there is accompanying overaction of the ipsilateral antagonist, the inferior oblique, so the affected eye elevates instead of depresses when put into this position.

Where an acquired fourth nerve palsy is suspected, an urgent referral (within one week) to orthoptics and ophthalmology should be instigated to confirm diagnosis and to complete further medical and neurological evaluations. If it is caused by a head trauma, it is important to rule out the possibility of a concurrent subarachnoid haemorrhage. If a longstanding, congenital fourth nerve palsy has recently decompensated, it is worthwhile referring urgently to rule out any sinister cause of decompensation, particularly in older patients (over 60 years) with an increased risk of vascular incident. Fresnel vertical prisms may be prescribed to alleviate the symptoms of diplopia or an occlusive patch or contact lens can be used until the condition resolves. Most vascular cases resolve within 3–6 months or at least, once stabilised, vertical prisms can be added permanently to the patient’s glasses to maintain binocularity. Where there is insufficient resolution and the deviation is too large to control with prisms, surgery may be considered.

About 75% of all fourth nerve palsies are congenital in origin and result either from dysgenesis of the fourth nerve itself, or from an abnormally long and lax superior oblique tendon. If detected in childhood, any significant refractive error should be prescribed and the child referred to orthoptics routinely to confirm diagnosis and for the management of any associated amblyopia. Often no further treatment is required if the child can maintain binocular single vision with a comfortable compensatory head posture.

**Summary**

Part 1 of this series has described the typical characteristics of incomitant strabismus and how a community optometrist should investigate if such a condition presents. Incomitant strabismus can occur from neurogenic, mechanical or myogenic causes or from letter pattern deviations; neurogenic causes and management have been discussed. Part 2 will cover the diagnosis and management of the other causes (mechanical, myogenic and letter pattern deviations). Optometrists should be able to differentially diagnose the types of incomitant strabismus that present to community practice using a range of common examination techniques. Once a differential diagnosis has been established, the optometrist should know the relevant optometric management options and appropriate referral guidelines to orthoptics and ophthalmology.

**References**

GOC’s Enhanced CET Scheme

CET and CPD regulators require practitioners to reflect on their learning. Additional activities are required to gain CET for distance learning.

Log into your CET dashboard via iLearn. On the menu reach you can choose either interactive or non-interactive CET for this unit of learning.

If you choose ‘non-interactive’, you have to pass (>60%) a six-question multiple-choice quiz. If you choose ‘interactive’, you must pass the MCQ quiz and complete a further 30-minute discussion with a colleague, and upload a short summary of your discussion and reflections within 30 days. Note you must complete both tasks before your CET can be awarded. If you want the CET counted within a calendar year, make sure you submit the online record of discussion and remind your colleague to verify it online at least 2 weeks before the end of the year.

Further instructions for interactive learning are as follows:

The following steps must be completed within 30 days of completing the MCQ quiz:

1. Discuss the interactive questions below with a registered colleague. Note if you are an optometrist, the colleague must also be an optometrist. If you are a dispensing optician, the colleague may be a dispensing optician, a contact lens optician or an optometrist. The discussion should be in a quiet environment where you are not interrupted for at least 30 minutes. Discuss the set questions and record a summary of the output of your discussion. Please ensure to create a paper copy of your record. Sign and date the document and keep it safely stored in case your CET is audited in future by the GOC.

2. In the event of an audit, we need to be able to show the GOC that the interaction has taken place in accordance with the instructions. Therefore, before you can be given points for this activity you must, within 30 days, record your answers to the set questions in the online Discussion Record and Reflection form (link provided on iLearn).

3. You will be asked for the GOC number, name and email address of the colleague who has completed the interaction with you, so please have those ready. Your colleague will be contacted by email (so please make sure you enter their correct email address) and will be sent a link to verify the interaction took place.

4. You can only be awarded interactive CET points if these steps are completed within 30 days.

The learning objectives for the article are:

8.1.1 Optometrists will have an enhanced understanding of characteristics of incomitant strabismus and the investigative procedures that aid differential diagnosis

8.1.4 Optometrists will have an enhanced understanding of the management options for incomitant strabismus of neurogenic origin and appropriate referral guidelines to orthoptics and ophthalmology.

2.2.2 Optometrists will have an enhanced understanding of the appropriate referral guidelines to orthoptics and ophthalmology for cases of incomitant strabismus of neurogenic origin

7.1.5 Dispensing opticians will have an enhanced understanding of the characteristics of incomitant strabismus, the investigative procedures to establish a diagnosis and management options for cases of neurogenic origin

The discussion tasks for the interactive learning option are as follows:

1. Discuss with your colleague the different characteristics of an incomitant strabismus.

2. Discuss with your colleague how to investigate an incomitant strabismus.

3. Discuss with your colleague the personal learning outcomes you have gained from this module and how you will apply this learning to practice. Consider the following questions (you will upload these reflections to iLearn and to myGOC within 30 days of completing the quiz).

   a. What are the main things you learned from the reading?
   b. How will you apply this learning in your future practice?
   c. Has this module identified any areas in which you wish to pursue further learning?