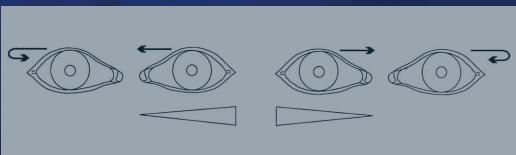


Clinical ORTHOPTICS

Fiona J. Rowe



THIRD EDITION



WILEY-BLACKWELL

Clinical Orthoptics

Dedication

This book is dedicated to my family

Clinical Orthoptics

Third Edition

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 WILEY-BLACKWELL

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Preface

Clinical Orthoptics has become established as a basic reference text providing fundamental information on anatomy, innervations and orthoptic investigation, plus diagnosis and management of strabismus, ocular motility and related visual disturbances. As with previous editions, the third edition is not designed to provide in-depth discussion of the content as it is recognised that this can be found in other excellent texts, in systematic reviews and in journal literature.

Following the revision of previous editions, this third edition, in addition to many of the original illustrations, contains new figures, tables and flowcharts designed to enhance the written text. Reference and further reading lists for each chapter have been extended and include up-to-date literature.

The layout of the text remains similar to that of the previous edition. Section I concentrates on anatomy and innervations of extraocular muscles including muscle pulley systems and associated cranial nerves. Ocular motility and orthoptic investigative techniques have been updated to include new assessments and reference to normative data. Section II refers to concomitant strabismus and Section III to incomitant strabismus. There has been considerable revision to add new information on conditions not previously included. A new chapter on craniofacial synostosis syndromes has been added. Section IV includes an updated list of abbreviations and glossary of definitions with additions to the information provided on diagnostic aids, flowcharts and illustrative case reports.

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Thanks are due to my colleagues and undergraduate students at the University of Liverpool, whose discussions provoke enquiry and understanding of orthoptics. Thanks are due to Addenbrooke's Hospital, Cambridge, for permission to use patient photographs and to the patients and parents for their consent to use these images. The glossary incorporates terminology from the British and Irish Orthoptic Society, and thanks are due to the Society for permission to use the glossary terminology. Finally, a thank you to the team at Wiley-Blackwell, the publisher, for their input to this text.

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SECTION I

1

Extraocular Muscle Anatomy and Innervation

This chapter outlines the anatomy of the extraocular muscles and their innervation and associated cranial nerves (II, V, VII and VIII).

There are four rectus and two oblique muscles attached to each eye. The rectus muscles originate from the Annulus of Zinn, which encircles the optic foramen and medial portion of the superior orbital fissure (Fig. 1.1). These muscles pass forward in the orbit and gradually diverge to form the orbital muscle cone. By means of a tendon, the muscles insert into the sclera anterior to the rotation centre of the globe (Fig. 1.2).

The extraocular muscles are striated muscles. They contain slow fibres, which produce a graded contracture on the exterior surface, and fast fibres, which produce rapid movements on the interior surface adjacent to the globe. The slow fibres contain a high content of mitochondria and oxidative enzymes. The fast fibres contain high amounts of glycogen and glycolytic enzymes and less oxidative enzymes than the slow fibres. The global layer of the extraocular muscles contains palisade endings in the myotendonous junctions, which are believed to act as sensory receptors. Signals from the palisade endings passing to the central nervous system may serve to maintain muscle tension (Ruskell 1999, Donaldson 2000).

Muscle pulleys

There is stereotypic occurrence of connective tissue septa within the orbit and stereotypic organisation of connective tissue around the extraocular muscles (Koornneef 1977, 1979). There is also stability of rectus extraocular muscle belly paths throughout the range of eye movement, and there is evidence for extraocular muscle path constraint by pulley attachment within the orbit (Miller 1989, Miller *et al.* 1993, Clark *et al.* 1999). High-resolution MRI has confirmed the presence of these attachments via connections that constrain the muscle paths during rotations of the globe (Demer 1995, Clark *et al.* 1997). CT and MRI scans have shown that the paths of the rectus muscles remain fixed relative to the orbital wall during excursions of the globe and even after large surgical transpositions (Demer *et al.*

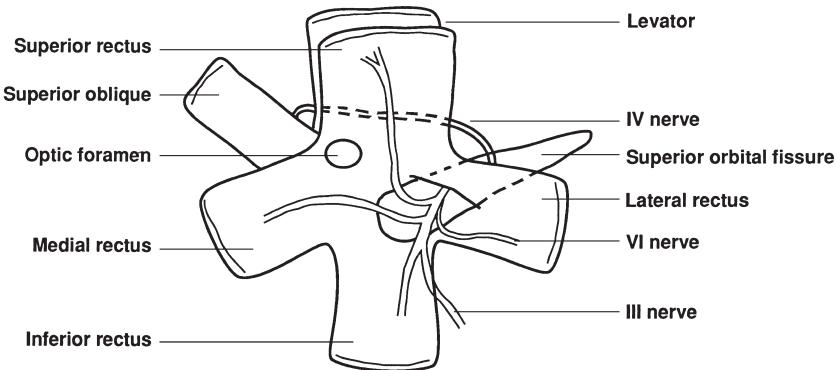


Figure 1.1 Orbital apex.

1996, Clark *et al.* 1999). It is only the anterior aspect of the muscle that moves with the globe relative to the orbit.

Histological studies have demonstrated that each rectus pulley consists of an encircling ring of collagen located near the globe equator in Tenon fascia attached to the orbital wall, adjacent extraocular muscles and equatorial Tenon fascia by sling-like bands, which consist of densely woven collagen, elastin and smooth muscle (Demer *et al.* 1995, Porter *et al.* 1996). The global layer of each rectus extraocular muscle, containing about half of all extraocular muscle fibres, passes through the pulley and becomes continuous with the tendon to insert on the globe. The orbital layer containing the remaining half of the extraocular muscle fibres inserts on the pulley and not on the globe (Demer *et al.* 2000, Oh *et al.* 2001, Hwan *et al.* 2007).

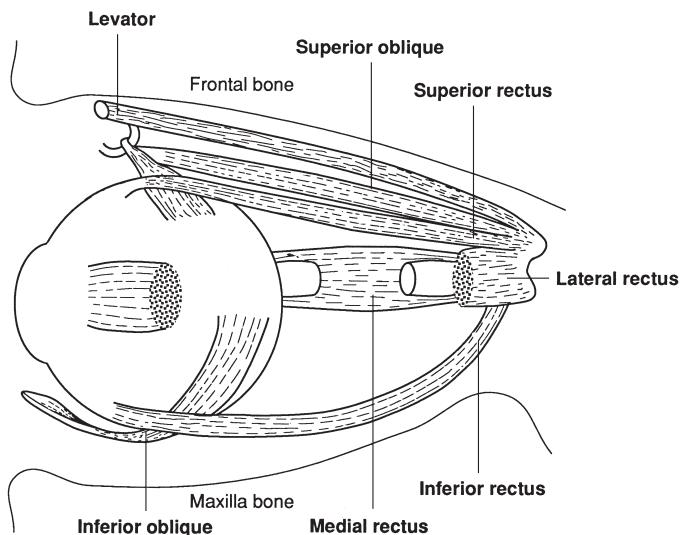


Figure 1.2 Extraocular muscles.

The orbital layer translates pulleys while the global layer rotates the globe through its insertion on the sclera. The inferior oblique muscle also has a pulley that is mechanically attached to the inferior rectus pulley (Demer *et al.* 1999).

The general arrangement of orbital connective tissues is uniform throughout the range of human age from foetal life to the tenth decade. Such uniformity supports the concept that pulleys and orbital connective tissues are important for the mechanical generation and maintenance of ocular movements (Kono *et al.* 2002).

Ocular muscles

Medial rectus muscle

This muscle originates at the orbital apex from the medial portion of the Annulus of Zinn in close contact with the optic nerve. It courses forward for approximately 40 mm along the medial aspect of the globe and penetrates Tenon's capsule roughly 12 mm from the insertion. The last 5 mm of the muscle are in contact with the eye and the insertion is at 5.5 mm from the limbus with a width of 10.5 mm. The muscle is innervated by the inferior division of the III nerve, which enters the muscle on its bulbar side. Its function is adduction of the eye (Fig. 1.3).

Lateral rectus muscle

This muscle arises by two heads from the upper and lower portions of the Annulus of Zinn where it bridges the superior orbital fissure. It courses forward for approximately 40 mm along the lateral aspect of the globe and crosses the inferior oblique insertion. It penetrates Tenon's capsule at roughly 15 mm from the insertion and the last 7–8 mm of the muscle is in contact with the eye. The insertion is at

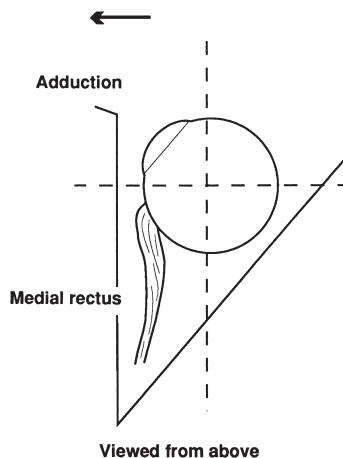


Figure 1.3 Medial rectus action.

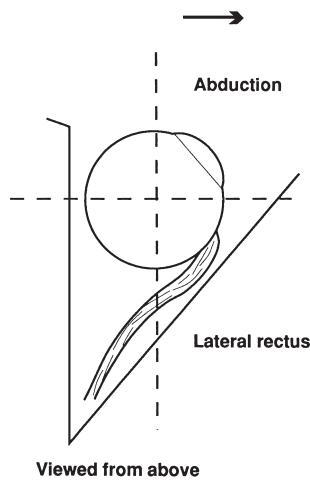


Figure 1.4 Lateral rectus action.

7 mm from the limbus with a width of 9.5 mm. The muscle is innervated by the VI nerve, which enters the muscle on its bulbar side. Its function is abduction of the eye (Fig. 1.4).

Superior rectus muscle

This muscle arises from the superior portion of the Annulus of Zinn and courses forward for approximately 42 mm along the dorsal aspect of the globe forming an angle of 23° with the sagittal axis of the globe. Superiorly, it is in close contact with the levator muscle. It penetrates Tenon's capsule at roughly 15 mm from the insertion and the last few mms of the muscle are in contact with the eye. The insertion is at 7.7 mm from the limbus with a width of 11 mm. The muscle is innervated by the superior division of the III nerve, which enters the muscle on its bulbar side. Its functions are elevation, intorsion and adduction of the eye (Fig. 1.5).

Inferior rectus muscle

This muscle arises from the inferior portion of the Annulus of Zinn and courses forward for approximately 42 mm along the ventral aspect of the globe forming an angle of 23° with the sagittal axis. It penetrates Tenon's capsule roughly 15 mm from the insertion and the last few millimetres of the muscle are in contact with the eye as it arcs to insert at 6.5 mm from the limbus. The width of insertion is 10 mm. The muscle is innervated by the inferior division of the III nerve, which enters the muscle on its bulbar side. Its functions are depression, extorsion and adduction of the eye (Fig. 1.6).

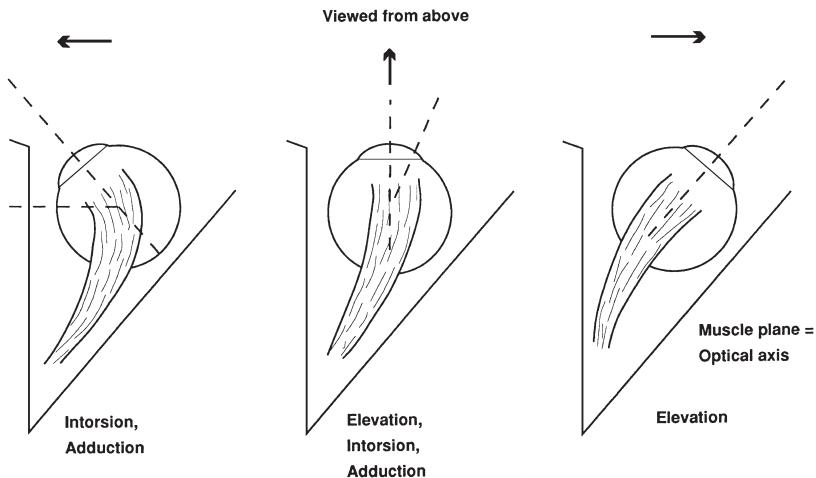


Figure 1.5 Superior rectus action. The course of the superior rectus is at an angle of 23° to the medial wall of the orbit. Actions in adduction are principally intorsion and adduction; in the primary position, actions are elevation, intorsion and adduction; action in abduction is principally elevation.

Superior oblique muscle

This muscle originates from the orbital apex from the periosteum of the body of the sphenoid bone, medial and superior to the optic foramen. It courses forward for approximately 40 mm along the medial wall of the orbit to the trochlea

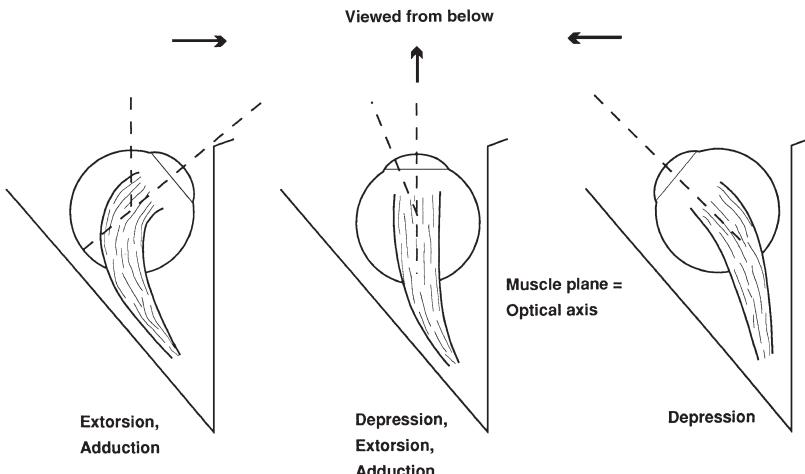


Figure 1.6 Inferior rectus action. The course of the inferior rectus is at an angle of 23° to the medial wall of the orbit. In adduction, the actions are principally extorsion and adduction; in the primary position, actions are depression, extorsion and adduction; action in abduction is principally depression.

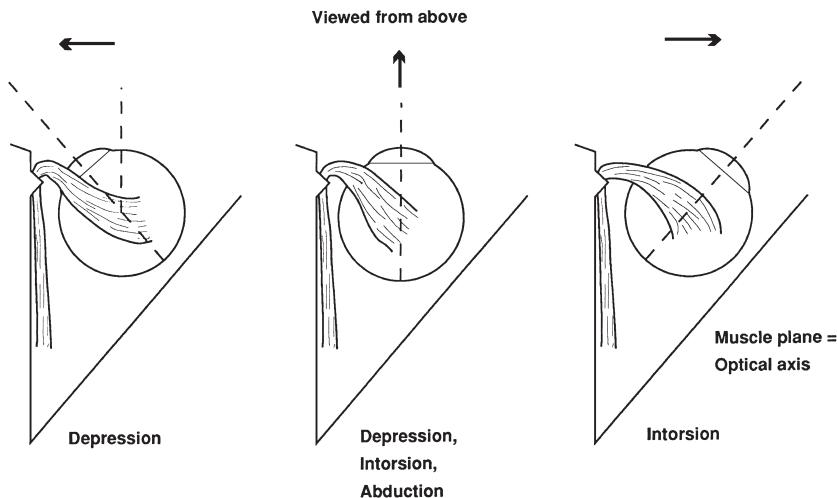


Figure 1.7 Superior oblique action. The course of the superior oblique tendon is at an angle of 51° to the medial wall of the orbit. Action in adduction is depression; in the primary position, actions are depression, intorsion and abduction; in abduction, action is intorsion.

(a V-shaped fibrocartilage that is attached to the frontal bone). The trochlear region is described by Helveston *et al.* (1982).

The muscle becomes tendonous roughly 10 mm posterior to the trochlea and is encased in a synovial sheath through the trochlea. From the trochlea, it courses posteriorly, laterally and downwards forming an angle of 51° with the visual axis of the eye in the primary position. It passes beneath the superior rectus and inserts on the upper temporal quadrant of the globe ventral to the superior rectus. Its insertion is fanned out in a curved line 10–12 mm in length. The muscle is innervated by the IV nerve that enters the muscle on its upper surface roughly 12 mm from its origin. Its functions are intorsion, depression and abduction of the eye (Fig. 1.7).

Inferior oblique muscle

This muscle arises from the floor of the orbit from the periosteum covering the anteromedial portion of the maxilla bone. It courses laterally and posteriorly for approximately 37 mm, forming an angle of 51° with the visual axis. It penetrates Tenon's capsule near the posterior ventral surface of the inferior rectus, crosses the inferior rectus and curves upwards around the globe to insert under the lateral rectus just anterior to the macular area. The muscle is innervated by the inferior division of the III nerve that enters the muscle on its bulbar surface. Its functions are extorsion, elevation and abduction of the eye (Fig. 1.8).

Figure 1.9 illustrates the muscle insertions in relation to the anterior segment of the eye. Figure 1.10 illustrates the positions of main action of each extraocular muscle and Table 1.1 illustrates all primary, secondary and tertiary muscle actions.

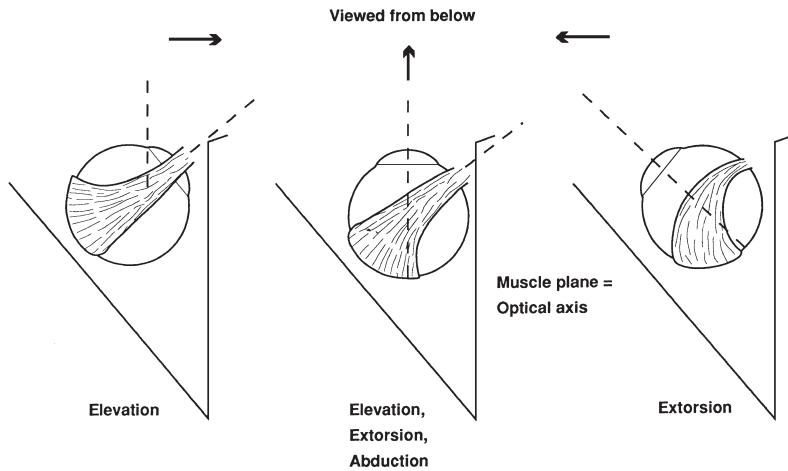


Figure 1.8 Inferior oblique action. The course of the inferior oblique is at an angle of 51° to the medial wall of the orbit. Action in adduction is elevation; actions in the primary position are elevation, extorsion and abduction; in abduction, action is extorsion.

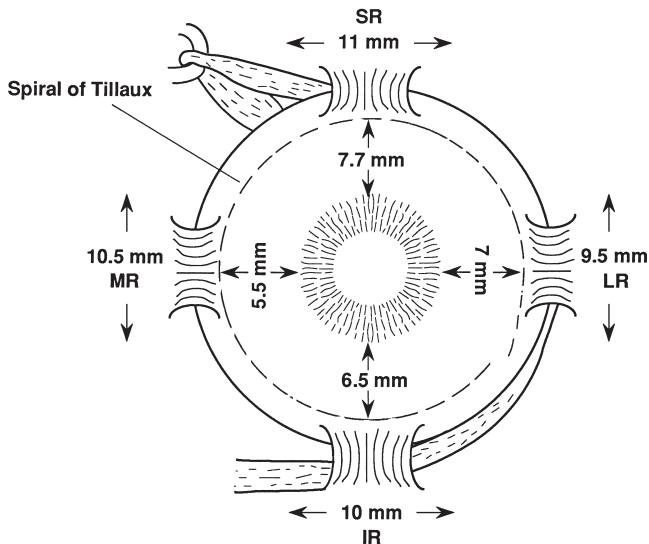


Figure 1.9 Extraocular muscle insertions. SR, superior rectus; MR, medial rectus; LR, lateral rectus; IR, inferior rectus.

SR	IO	IO	SR
↖	↗	↖	↗
LR	↖ RIGHT	↗ MR	↖ LEFT
↖	↘	↖	↘
IR	SO	SO	IR
SR Superior rectus	IR Inferior rectus	LR Lateral rectus	
MR Medial rectus	IO Inferior oblique	SO Superior oblique	

Figure 1.10 Cardinal positions of gaze – position of main action of extraocular muscles.

Table 1.1 Primary, secondary and tertiary extraocular muscle actions.

Muscle	Primary action	Secondary action	Tertiary action
Medial rectus	Adduction	–	–
Lateral rectus	Abduction	–	–
Superior rectus	Elevation, maximum in abduction	Intorsion, maximum in adduction	Adduction, maximum in adduction
Inferior rectus	Depression, maximum in abduction	Extorsion, maximum in adduction	Adduction, maximum in adduction
Superior oblique	Intorsion, maximum in adduction	Depression, maximum in abduction	Abduction, maximum in abduction
Inferior oblique	Extorsion, maximum in abduction	Elevation, maximum in adduction	Abduction, maximum in abduction

Levator palpebral superioris

This muscle originates from the under surface of the lesser wing of sphenoid bone above and in front of the optic foramen by a short tendon that blends with the origin of the superior rectus. It runs forward and changes directly from horizontal to vertical at the level of the equator of the globe. At approximately 10 mm above the superior margin of the tarsus, it divides into anterior and posterior lamellae. The anterior lamellae form the levator aponeurosis that is inserted into the lower third of the entire length of the anterior surface of the tarsus. Its fibres extend to the pretarsal portion of the orbit and skin. The posterior lamellae form Muller's muscle that is attached inferiorly to the superior margin of the tarsus.

Innervation

The extraocular muscles are innervated by the III, IV and VI nerves.

III nerve

The III nerve (third/oculomotor) supplies the superior rectus, inferior rectus, medial rectus, inferior oblique and levator muscles. Its visceral fibres innervate the ciliary muscle and sphincter pupillary muscle that synapse in the ciliary ganglion.

The nuclei are in the mesencephalon at the level of the superior colliculus. There is an elongated mass of cells that form the nuclei. Peripheral motor neurones innervate multiply innervated extraocular muscle fibres and central motor neurones innervate single innervated muscle fibres. Dorsal nucleus fibres pass to the ipsilateral inferior rectus, intermediate nucleus fibres pass to the ipsilateral inferior oblique, ventral nucleus fibres pass to the ipsilateral medial rectus, paramedian nucleus fibres pass to the contralateral superior rectus, central caudal nucleus fibres pass to both levator muscles, and the anterior median/Edinger-Westphal nucleus

contains the parasympathetic fibres (Bienfang 1975). The nerve fibres emerge from the mesencephalon ventrally where they are closely associated with the posterior cerebellar and superior cerebral arteries. The nerve courses forward through the subarachnoid space to pierce the dura mater at the posterior clinoid process and enter the cavernous sinus.

The third cranial nerve pathway is supplied by branches of the basilar artery including the superior cerebellar arteries, posterior cerebral arteries, mesencephalic perforating arteries, collicular and accessory arteries in the midbrain; the thalamoperforating arteries supplemented by the superior cerebellar artery, posterior communicating artery and posterior cerebral artery in the proximal nerve pathway; and inferior cavernous sinus arteries, medial posterior choroidal artery and tentorial arteries in the distal nerve pathway (Marinkovic & Gibo 1994, Cahill *et al.* 1996).

IV nerve

The IV nerve (fourth/trochlear) supplies the superior oblique. The nucleus lies in the mesencephalon at the level of the inferior colliculus. The nerve fibres decussate (although about 3% do not decussate but retain ipsilateral projection) and emerge from the brainstem dorsally. The nerves curve around the brainstem and course forward through the subarachnoid space to pierce the dura mater and enter the cavernous sinus.

The fourth cranial nerve pathway is in close association or contact with branches of the basilar artery in the midbrain including the superior cerebellar artery, vernian artery and collicular artery. It is supplied by posterior cerebral artery and posterior communicating artery in its proximal pathway and by the internal carotid artery, medial posterior choroidal artery and tentorial arteries in the distal pathway (Marinkovic *et al.* 1996, Yousry *et al.* 2002).

VI nerve

The VI nerve (sixth/abducens) supplies the lateral rectus. The nucleus is situated in the pons in the floor of the IV ventricle near the midline, medial to VIII nucleus and proximal to the paramedian pontine reticular formation. The medial longitudinal fasciculus lies medial to the nucleus. The nerve fibres emerge from the brainstem ventrally and course forward and laterally over the petrous tip of the temporal bone and under the petrosphenoid ligament. The nerve pierces the dura mater to enter cavernous sinus. The nerve divides into two distinct trunks along its pathway between the brainstem and the lateral rectus muscle.

The sixth cranial nerve pathway is supplied with branches of the basilar artery including the anterior inferior cerebellar artery, posterior inferior cerebellar artery, pontomedullary artery and accessory arteries in the pons and clivus region. The distal pathway is supplied by the internal auditory artery, anterolateral artery and tentorial artery (Marinkovic *et al.* 1994, Yousry *et al.* 1999).

Common nerve pathways

The III, IV and VI nerves course forward together in the lateral aspect of the cavernous sinus entering the orbit through the superior orbital fissure. The III and VI nerves enter within the muscle cone.

The III nerve divides into the superior and inferior divisions. The superior division enters the superior rectus on its bulbar surface and passes through the muscle to terminate in the levator muscle. The inferior branch supplies the medial rectus, inferior rectus, and then passes beneath the optic nerve to the floor of the orbit and terminates in the inferior oblique. The terminal branch also sends a short branch to the ciliary ganglion. The VI nerve passes forward and laterally to enter the lateral rectus bulbar surface. The IV nerve enters through the superior orbital fissure laterally and superior to the Annulus of Zinn. It passes anteriorly and medially crossing the III nerve, levator muscle and superior rectus, and enters the superior oblique on its orbital surface.

Associated cranial nerves

Autonomic nerves

These nerves supply smooth muscles and source ganglia. Smooth muscles include the muscular blood vessels, Muller's muscle, pulley smooth muscle, sclera myofibroblasts and choroidal smooth muscle (Demer *et al.* 1997). Source ganglia include the pterygopalatine ganglion, ciliary ganglion and superior cervical ganglion.

Proprioceptive nerves

These nerves consist of palisade endings and spindles. Palisade endings innervate myotendonous cylinders at the termination of each multiply innervated global layer fibre in the rectus extraocular muscles (Lienbacher *et al.* 2011). Spindles are composed of several orbital layer myofibres and have nerve terminals within a very thin capsule.

II nerve

The II (optic) nerve serves the sensory function of vision. Its pathway commences in the eye at the receptor cells in the retina. There is a complex arrangement of nuclei and processes from three layers of photoreceptors, bipolar cells and ganglion cells. There are in the region of 1.2–1.5 million retinal ganglion cells and 105 million photoreceptors with an average ratio of 1 retinal ganglion cell to 100 photoreceptors. At the fovea, the ratio is 1:1 for retinal ganglion cells to photoreceptors.

Retinal ganglion cells include midget (parvocellular), parasol (magnocellular), koniocellular and other cells. Midget ganglion cells are responsible for slow conduction of impulses with low temporal resolution and require high contrast

stimuli. Parasol ganglion cells are responsible for fast conduction of impulses with high temporal resolution and requiring low contrast stimuli. Midget cells have colour selectivity whereas parasol cells have little or no colour selectivity. Konio-cellular cells have moderate conduction velocity and moderate sensitivity to light and spatial resolution. They have some colour selectivity and may have a role in motion detection and visual attention. Other cells include light reflex ganglions and photosensitive neurones.

Retinal ganglion cells pass in nerve fibre bundles to the optic discs and pass from each eye to the intracranial cavity along the optic nerves. The optic nerves merge in the optic chiasm where there is crossing of nasal retinal fibres. Ipsilateral temporal and contralateral nasal fibres pass along the optic tracts to the lateral geniculate nuclei where the first synapse of retinal nerve fibres occurs. The post-synaptic fibres then pass via the optic radiations to the visual cortex. The visual cortex (V1) occupies the calcarine sulcus in the occipital lobe and is the primary visual area.

V nerve

The V nerve (fifth/trigeminal) serves sensory and motor functions and the nuclei extend through the pons down into the medulla. The sensory nerve has three branches.

Sensory nerves

The ophthalmic division serves the sensory function to the lacrimal gland, conjunctiva, forehead, eyelids, anterior scalp and mucous membranes of the nose. The sensory fibres pass through the superior orbital fissure to the cavernous sinus and pass inferiorly to the trigeminal ganglion, which is located under the cavernous sinus in Meckel's cave (a groove in the skull). Fibres pass from the ganglion posteriorly to the pons to the trigeminal nuclei.

The maxillary division serves the sensory function to the cheeks, upper gums and teeth and lower eyelids. The sensory fibres pass through the foramen rotundum, underneath the cavernous sinus to the trigeminal ganglion and then onto the nuclei in the pons.

The mandibular division serves the sensory function to the teeth, gums of the lower jaw, pinna of ears, lower lip and tongue. The sensory fibres pass through the foramen ovale underneath the cavernous sinus to the trigeminal ganglion and then onto the nuclei in the pons.

Motor nerves

Motor fibres of the V nerve serve the muscles of mastication. The motor nuclei are located in the pons near the seventh nerve nuclei and aqueduct. Nerve fibres leave ventrally and medially and pass anteriorly to the trigeminal ganglion, through the foramen ovale to the muscles of mastication.

VII nerve

The VII nerve (seventh/facial) serves sensory and motor functions. The VII nerve has central connections to the motor face area of the cerebral cortex and the nuclei are divided into upper and lower halves. Corticobulbar fibres double decussate for the upper face but there is single decussation for lower face fibres.

Sensory fibres

Ganglion cells supply taste buds in the palate and tongue and sensory fibres are also present in the skin, in and around the external acoustic meatus. Fibres pass to the geniculate ganglion situated in the internal auditory meatus and pass back to the pons.

Motor fibres

The nuclei are located in the lateral part of the pons and fibres loop around the abducens nuclei, forming the facial colliculus, before leaving the pons ventrally. Fibres pass anteriorly and enter the internal auditory meatus. The nerve enters a narrow bony canal above the labyrinth and descends to the stylomastoid foramen where a branch supplies the stapedius muscle. It leaves the skull and supplies the facial muscles (frontal, zygomatic, buccal, mandibular marginal and cervical branches).

VIII nerve

The VIII nerve (eight/auditory) serves the sensory function of hearing and balance.

Cochlear nerve (hearing)

Receptor cells are hair cells in the organ of Corti. Fibres pass from the spiral ganglion along the Cochlear nerve through the internal auditory meatus to the cisterna pontis, to the inferior cerebellar peduncle and to the cochlear vestibular nuclei in the pons/medulla.

Vestibular nerve (balance)

Receptor cells are hair cells in the utricles, saccules and semicircular canals. Fibres pass from Scarpa's ganglion along the vestibular nerve through the internal auditory meatus to the cisterna pontis and to the vestibular nuclei in the pons/medulla. Within the internal auditory meatus, the vestibular and cochlear nerves are in close association with the facial nerve. Within the acoustic foramen and intracranial cavity, these nerves are closely associated with both the sixth and facial nerves.

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2

Binocular Single Vision

Binocular single vision is the ability to use both eyes simultaneously so that each eye contributes to a common single perception.

Normal binocular single vision occurs with bifoveal fixation and normal retinal correspondence in everyday sight. Abnormal binocular single vision occurs in the absence of bifoveal fixation usually with abnormal retinal correspondence in everyday sight.

Worth's classification

Binocular single vision can be classified into three stages:

1. Simultaneous perception and superimposition
2. Fusion
3. Stereoscopic vision

Simultaneous perception is the ability to perceive simultaneously two images, one formed on each retina. Superimposition is the simultaneous perception of the two images formed on corresponding areas, with the projection of these images to the same position in space. This may occur whether the correspondence is normal or abnormal. If fusion is absent, two similar images are seen as separate but superimposed and no fusion range is demonstrable.

Fusion may be sensory or motor. Sensory fusion is the ability to perceive two similar images, one formed on each retina, and interpret them as one. Motor fusion is the ability to maintain sensory fusion through a range of vergence, which may be horizontal, vertical or cyclovergence. Stereoscopic vision is the perception of the relative depth of objects on the basis of binocular disparity.

Development

The initial ocular position in the human neonate (Rethy 1969) is often one of divergence. In the early postnatal period, divergence decreases towards a binocular

coincidental position resulting in similar visual stimulation of each eye, which in turn facilitates firing of binocular driven cells/neurones in the striate cortex (Hubel & Wiesel 1968), and once the straight position is attained, this is maintained preferentially. The globes are approximately 73% of adult size in infancy and the lens is relatively less convex. The size of eye (lens and cornea) renders the eye hypermetropic, which is overcome by amplitude of accommodation. Other developments include accommodation, ocular muscles and nerve supply, visual pathway and foveal development. Binocular single vision is dependent on retinal correspondence and disparity. Eighty percent of the striate cortex neurones are driven from either eye (binocular driven cells) whilst 10% are driven by the right eye only and 10% by the left eye only (Hubel & Wiesel 1968).

Some sensory and motor binocular associations exist in the visual system of the newborn. The binocular reflexes relate to the development of binocular single vision on the basis of continued use of the visual system. Postural reflexes are inborn and must be present if binocular single vision is to develop:

- a. Static reflexes compensate for changes in position of the head relative to the body.
- b. Statokinetic reflexes compensate for changes in head position relative to space.

Fixation reflexes form the mechanism from which binocular vision develops:

- a. The primary fixation reflex achieves foveal fixation in either eye and is present at birth.
- b. The refixation reflex allows foveal refixation from target to target and maintenance of foveal fixation on a moving target. This develops from 6 to 8 weeks of age. Most neonates are capable of locating and briefly fixing a moving target, and the eyes can move in a coarsely conjugate fashion. Thus, the refixation reflex, although unstable, appears to be present at birth, despite the fact that in the first months of life the fovea is still poorly differentiated.
- c. The conjugate fixation reflex, where the eyes learn to move together during versions, is the first reflex by which the eyes move binocularly and develops from 2 to 3 weeks of age.
- d. The disjunctive fixation reflex allows binocular vision to be maintained through the range of vergence movements that follow changes of fixation distance and develops from 2 to 3 months of age.
- e. The corrective fusion reflex allows binocular vision to be maintained under conditions of stress, such as overcoming prisms in clinical testing situations, and develops from 3 to 6 months of age.
- f. Kinetic reflexes maintain binocular single vision through controlled accommodation and convergence.

The newborn does not converge the eyes, but the attempt to converge may be seen as early as 1 month after birth. The macula is poorly developed at birth with incomplete migration of retinal ganglion cells from the foveal area. Saccadic eye

movements are poorly controlled, and several movements are required to achieve foveation. By 5–6 weeks of age, the conjugate fixation reflex is developed, and the two eyes conjugately fix an object and follow it over a considerable range for at least a few seconds. Density of cones in the fovea increases with myelination of nerve fibres in the visual pathway. Smooth pursuit eye movement and colour vision develop from 2 months. Accommodation develops rapidly from 2–3 months and approaches the same levels of accuracy as adults (Horwood & Riddell 2004). Optokinetic nystagmus is asymmetrical up to 3 months with absent nasal to temporal movement, after which there is progression to symmetrical motion processing by the age of 6 months (Bosworth & Birch 2007). By 6 months, the conjugate movements of binocular vision become accurate and convergence is well developed. By 6–8 months, a fusional movement can be detected by placing a small prism over either eye.

A critical period occurs in the development of the visual system during which the visual system is susceptible to abnormal visual input (Daw 1998). The period of normal visual development is up to 5 years of age.

Retinal correspondence

This concerns the retinal areas of each eye that have the same visual direction during binocular vision.

Normal retinal correspondence

This is a binocular condition in which the fovea and areas on the nasal and temporal side of one retina correspond to and have, respectively, common visual directional sensitivity with the fovea and temporal and nasal areas of the retina of the other eye. Normal retinal correspondence is the normal state in which the visual direction of each fovea is the same (Flom & Weymouth 1961, Flom & Kerr 1967) (Fig. 2.1).

Abnormal retinal correspondence

This is a binocular condition in which there is a change in visual directional sensitivity such that the fovea of the fixing eye has a common visual directional sensitivity with an area other than the fovea of the deviating eye (Burian 1951). The pairing of all retinal areas is similarly changed. The condition may occur whichever eye is used for fixation (Fig. 2.2).

Harmonious abnormal retinal correspondence is present where the angle of anomaly is equal to the objective angle, and the subjective angle is zero. Unharmonious abnormal retinal correspondence is present where the angle of anomaly is different from the objective angle. The angle of anomaly is the difference between the objective and subjective angles of deviation. Abnormal retinal correspondence is present in constant manifest strabismus usually of a small angle less than 20 prism dioptres.

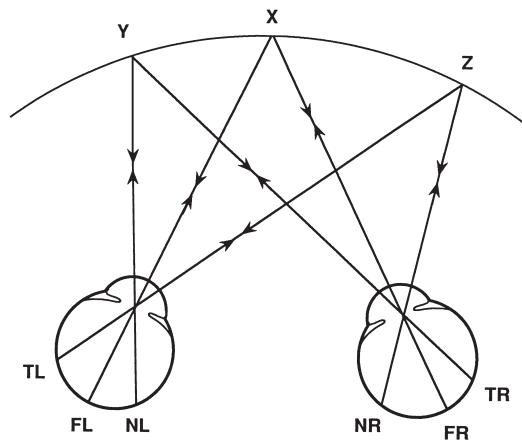


Figure 2.1 Projection in normal retinal correspondence with normal binocular single vision. Stimulation of corresponding points in both eyes results in localisation of the stimulus in the same direction in space. Both fovea, FL and FR, are corresponding points as are points on temporal and nasal retinas, TL and NR, NL and TR.

Physiology of stereopsis

The locations of all points in space that are imaged on corresponding retinal points are termed the *horopter*. Panum's space is a narrow band around the horopter within which object points give rise to binocular single vision. Objects are seen as single even though the object stimulates slightly disparate retinal elements.

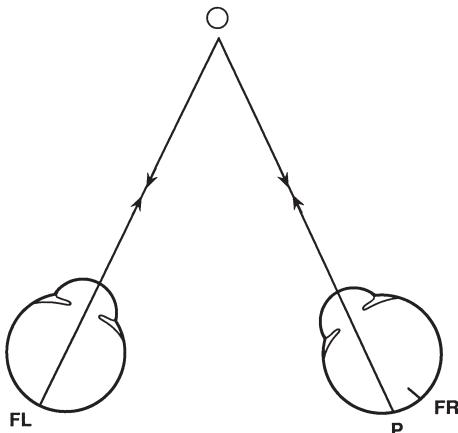


Figure 2.2 Projection in abnormal retinal correspondence with right esotropia. O is the fixation target. FL and FR are the fovea of both eyes and P is the pseudofovea of the right eye. FL and P are corresponding points as the right eye undergoes sensory adaptation with abnormal retinal correspondence. Stimulation of these corresponding points results in a single perception of the fixation target.

Panum's area is the retinal area surrounding one corresponding retinal point within which disparity of correspondence may occur, whilst maintaining binocular single vision. Binocular single vision is the result not of a rigid point-to-point correspondence but of a point-to-area relationship. The amount of foveal image disparity that permits fusion is small, and disparity increases gradually from the fovea to the periphery. Panum's area is narrow at the fixation point and widens towards the periphery. The horizontal area at the fovea is approximately 6–10 minutes, and this increases towards the periphery measuring approximately 30–40 minutes at 12° from the fovea. It may be larger than this as moving random-dot stereograms have shown fusion of disparities of 2°–3° (Hyson *et al.* 1983, Erkelens & Collewijn 1985, Piantanida 1986). Increases can be related to anatomical and physiological differences known to exist between the foveal cone system and the rod and cone system of the peripheral retina. The increase in Panum's area parallels the increase in size of the retinal receptive fields. Performance with motor skills is related to the level of stereoacluity in that performance is considerably worse in the absence of stereopsis (O'Connor *et al.* 2010).

Physiological diplopia

This is a type of diplopia that exists in the presence of binocular vision. It consists of the appreciation that a near object appears double when a distant object is fixated (heteronymous or crossed diplopia), and a distant object appears double when a near object is fixated (homonymous or uncrossed diplopia) (Figs. 2.3 and 2.4).

All objects outside Panum's space give rise to physiological diplopia. Physiological diplopia indicates that the patient is capable of using both eyes and is not suppressing one eye.

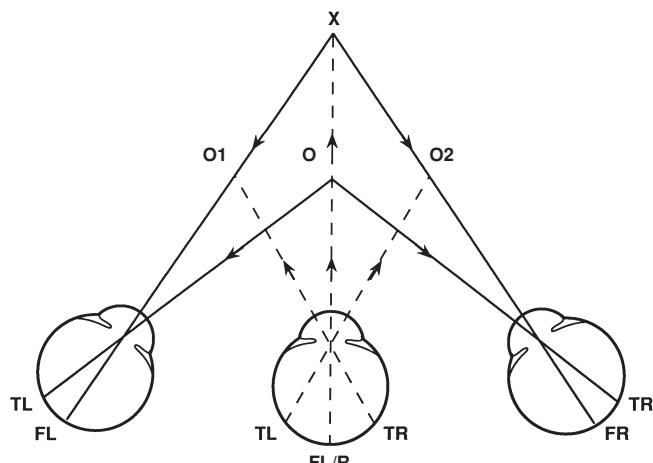


Figure 2.3 Projection in heteronymous diplopia. X is the fixation target, which is seen as a single image by the corresponding fovea FL and FR. O is an object between the fixation target and the eyes that stimulates non-corresponding temporal retinal points, TL and TR, resulting in crossed physiological diplopia.

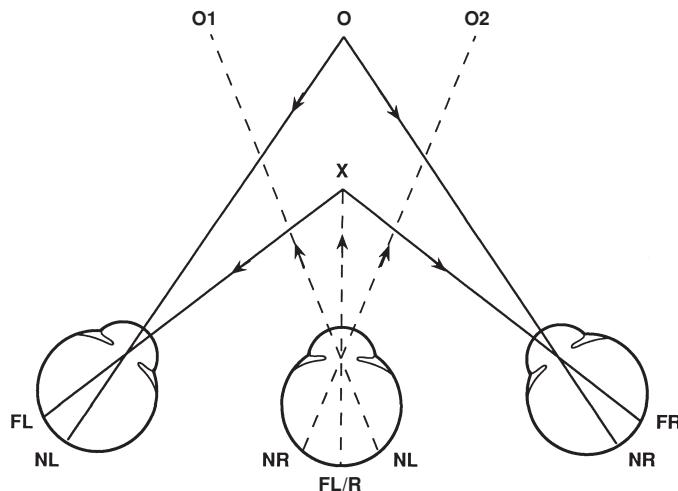


Figure 2.4 Projection in homonymous diplopia. X is the fixation target seen as a single image by the corresponding foveal points, FL and FR. O is an object further away than the fixation target that stimulates non-corresponding nasal retinal points, NL and NR, resulting in uncrossed physiological diplopia.

Fixation disparity

Fixation disparity is a phenomenon that occurs in binocular single vision in which the image is seen singly despite a slight underconvergence or overconvergence of the visual axes; the fixation target is imaged on slightly disparate retinal points within Panum's area. There is an apparent displacement of unioocularly observed details of targets whose other details are fused binocularly.

The phenomenon can be demonstrated clinically when targets, which have mainly identical features but also contain some dissimilar features, are presented to the eyes. Fusion occurs for the identical features, but a displacement occurs for the dissimilar features in the direction of the projection of the existing heterophoria.

Fixation disparity may be involved in the maintenance of binocular single vision. Disparity of retinal images causes fusional movements. At the end of a fusional movement, not all the disparity is annulled; a small disparity remains, which acts as an error signal. The residual fixation disparity may control the direction and strength of the innervation that maintains the new binocular position.

When visual objects are fused by being imaged on horizontally disparate points, within Panum's space, stereopsis results. The greater the horizontal disparity, the greater the depth effect. A vertical disparity produces no stereoscopic effect (Fig. 2.5).

Local stereopsis occurs where localised features of objects are extracted from a visual scene and assigned relative depth values, indicating that one feature is further away from another. Global stereopsis occurs where the perception of whole objects in stereoscopic depth is achieved.

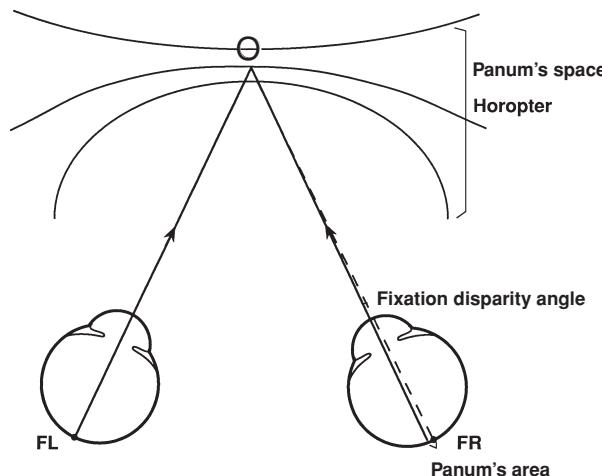


Figure 2.5 Projection in stereopsis. O is the fixation target, which projects directly to the fovea of the left eye. This should also project directly to the right eye. However, due to slight under- or overconvergence of the right eye, direct stimulation may not occur, resulting in fixation disparity. In this figure, there is slight overconvergence of the right eye with stimulation of a point just nasal to the fovea. As this point falls within Panum's area, the stimuli to either eye are fused, resulting in a single image of the fixation target.

Monocular clues are important in the estimation of the relative distance of visual objects and are active in monocular as well as binocular vision. These clues are the result of experience:

- *Motion parallax*: Targets further away move more in the horizontal plane than nearer targets.
- *Linear perspective*: Objects of the same size decrease in size the further away they are.
- *Overlapping contours*: Overlap in images determines which is in front or behind the other.
- *Distribution of highlights and shadows*.
- *Size of known objects*: When size is known, the relative distance can be determined.
- *Aerial perspective*: Knowledge of colours and hues.

Fusion

Central fusion occurs when the images of an object are perceived by each fovea, and the area surrounding them, and are unified. This produces bifoveal binocular single vision with fusion. The highest levels of stereoacuity are associated with central fusion. Peripheral fusion results from unification of images outside the central region. Gross stereopsis is associated with peripheral fusion. Central and peripheral fusion usually function simultaneously. A patient's sensory status is considered crucial to

the long-term stability of a successful surgical outcome. Fusion serves as the glue to maintain alignment (Burian 1941, Kushner & Morton 1992, Morris *et al.* 1993). It indicates the patient's ability to control their latent tendency for the eyes to drift. In order for fusion to occur, the images presented to each eye must be similar in size, brightness and sharpness. Peripheral fusion contributes significantly to the maintenance of binocular single vision (Bielchowsky 1935). If this is destroyed, even while maintaining good central vision, disruption of binocularity occurs.

When measured with large field stimuli, 8° of motor cyclovergence has been demonstrated in normals (Guyton 1988). Therefore, individuals can use this ability to fuse torched images without diplopia. This ability derives from the receptive fields in the peripheral retina being large compared to those in the central retina. This amount of motor cyclovergence combined with the 8° of sensory cyclofusion allows norms to fuse up to 16° of cyclodisparity.

Retinal rivalry

When dissimilar images are presented to corresponding retinal areas, fusion becomes impossible and retinal rivalry occurs. When dissimilar targets are presented to each eye, the patient will see one target, then the other, or a mosaic of contours, but not both simultaneously. Retinal rivalry is a physiological finding in binocular single vision and is distinct from suppression as it indicates a state of fluctuation between competing components. Retinal rivalry may also be produced by differences in colour and unequal illumination.

Suppression

Suppression is the mental inhibition of visual sensations of one eye in favour of those of the other eye when both eyes are open. This may occur in binocular single vision and commonly in manifest strabismus.

Physiological suppression is present in binocular single vision. Blurred images are suppressed when concentrating on one particular object. Pathological suppression is present in manifest strabismus and may alternate with alternating deviations (Fig. 2.6).

Suppression may occur with interocular blur, suspension, binocular retinal rivalry or permanent suppression. Interocular blur arises where there is a significant difference in blur or contrast between the two eyes such as with anisometropia, unequal amplitude of accommodation or asymmetric accommodation. Suspension relates to physiological suppression during physiological diplopia. Binocular retinal rivalry with differences from either eye in object size or shape prevents fusion. This can be exclusively dominant, in which one image swaps with the other and back; mosaic dominant, in which small interwoven retinal patches alternate; or luminance, which involves colour rivalry. Permanent suppression occurs where the individual is unable to see the object. Suppression occurs at a cortical level and may involve inhibitory interaction between neighbouring ocular dominance columns (Sengpiel *et al.* 1994).

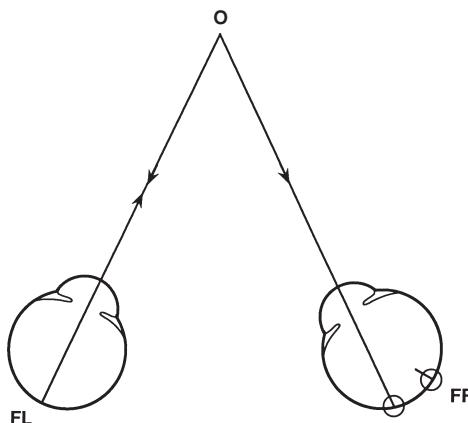


Figure 2.6 Projection in right esotropia with suppression. O is the fixation target, which stimulates the fovea of the left eye and a nasal retinal point of the right eye. The nasal retinal point is suppressed, which ensures a single perception of the fixation target.

The area and density of suppression will vary according to the type of strabismus (Harrad 1996). A defined small central suppression scotoma exists in microtropia whereas larger angle strabismus shows an elliptical shaped scotoma extending horizontally, particularly in esotropia. Suppression can be more extensive involving the entire hemifield as has been reported in exotropia (Jampolsky 1955).

Suppression typically develops in childhood strabismus. There is an issue as to whether adults can truly develop suppression. Inattention to diplopia and true suppression are different, but probably related, adaptive sensory strategies and additive factors other than the age of the patient may be involved (McIntyre & Fells 1996). Development of 'suppression' has been reported in adults with thyroid eye disease (Fells 1979), following retinal detachment surgery (Wright *et al.* 1999) and keratoconus (Sherafat *et al.* 2001). In adult onset strabismus, patients are seen to have a poor prognosis with regard to the ability to learn to suppress or ignore diplopia. Patients can have persistent troublesome diplopia that can be difficult or impossible to alleviate. An ability to develop suppression as an adult or an inattention to the diplopic image may reflect an element of plasticity in the mature binocular visual system.

Diplopia and confusion are not appreciated where suppression is present. Confusion is the simultaneous appreciation of two superimposed images due to the stimulation of corresponding retinal points by two different images. Binocular single vision is not present with pathological suppression, and suppression obstructs attempts to obtain binocular single vision.

Diplopia

Pathological binocular diplopia results from the presence of a manifest ocular deviation and is the simultaneous appreciation of two separate images caused by

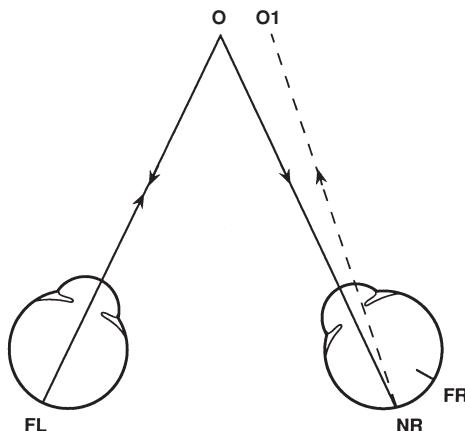


Figure 2.7 Projection in right esotropia with normal retinal correspondence and pathological diplopia. O is the fixation target, which stimulates non-corresponding points of the left fovea and right nasal retina. The nasal retina projects temporally, resulting in uncrossed pathological diplopia.

the stimulation of non-corresponding points by one object. It may be horizontal, vertical or torsional, or any combination.

Heteronymous diplopia is crossed binocular diplopia associated with exotropia in which the image of the fixation object is received on the temporal area of the retina of the deviating eye and is projected nasally. Homonymous diplopia is uncrossed binocular diplopia associated with esotropia in which the image of the

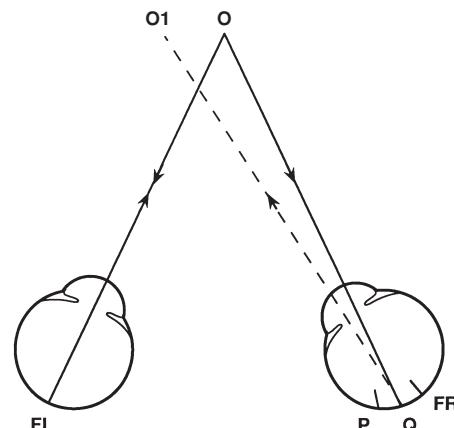


Figure 2.8 Projection in right esotropia with paradoxical diplopia. With abnormal retinal correspondence, there is sensory adaptation of the strabismic eye such that the pseudofovea and the fovea of the other eye become corresponding points. The retinal area between the fovea and pseudofovea of the strabismic eye, which was originally nasal retina, takes on the sensory function of temporal retina. With redirection of the strabismic eye, non-corresponding points are stimulated (FL and Q), resulting in crossed paradoxical diplopia.

fixation object is received on the nasal area of the retina of the deviating eye and is projected temporally (Fig. 2.7).

Paradoxical diplopia is pathological binocular diplopia in which heteronymous diplopia occurs in esotropia or homonymous diplopia occurs in exotropia (Fig. 2.8).

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3 Ocular Motility

A number of systems are responsible for producing eye movements and controlling fast (saccades), slow (smooth pursuit), disjunctive (vergence) and involuntary (vestibular) movements. The eye movement system used depends on the nature of the visual input received and the response required for the visual stimulus. Visual information reaches the visual striate cortex (primary visual area 17, V1) via the visual pathway and attributes of this visual information such as fine features, colour, spatial localisation and motion are processed in separate secondary visual areas (Rowe 2003).

Saccadic system

Saccadic eye movements are fast, ballistic eye movements under both volitional and reflex control with a maximum velocity of up to $500^{\circ}/\text{s}$. Voluntary movements include willed refixations, those in response to command and memory-guided saccades. Reflex movements include saccades in the direction of a new stimulus that may be visually guided (prosaccade) or in response to an auditory stimulus. These saccades may be accompanied by head movement in the same direction. The visual stimulus for generation of saccadic eye movement is target displacement in space. Control decisions of movement size and velocity are made on the basis of continuous inflow of visual information from the retina (Rowe 2003). Saccades are under an adaptive control system in order to maintain binocular control and to compensate for short- or long-term changes to the visual system. This adaptive control monitors performance and adjusts parameters to improve accuracy and behaviour where required. A learning process is required for saccadic adaptation due to the nature of saccades being ballistic rendering ongoing feedback impossible (Griffiths 2007).

The latency period is the interval from the appearance of a stimulus to the start of the eye movement in response. The ocular motor system for saccades responds after a latency or delay of 100–300 ms. Peak velocity and duration of saccade are dependent on size or amplitude of eye movement. Latencies are less when the

fixation target is turned off before the peripheral target appears (gap stimulus) and greater when the fixation target remains illuminated after the peripheral target appears (overlap stimulus) (Kalesnykes & Hallett 1987).

The saccadic response consists of a period of acceleration to peak velocity followed by deceleration of the eyes as they approach the target position. Extraocular muscles use relative tension to hold the new eye position along with check ligaments and fatty tissue of the orbit. Horizontal saccades are naturally disconjugate resulting in faster and slightly large abducting saccades in comparison to adducting saccades (Collewijn *et al.* 1988).

A pulse-step mechanism is the process that generates a saccadic eye movement (Optican & Miles 1985). Burst and pause cells are the critical components of this mechanism (Sparks & Mays 1990, Enderle 2002). The saccadic pulse is a burst of activity that generates the forces necessary to overcome the drag from orbital tissue content and produce eye movement. The number of spikes in a burst is proportional to the amplitude of the saccade and firing rate is proportional to eye velocity. Burst cells fire while omnipause cells inhibit movement thus constituting the pulse signal. Saccadic eye movement occurs and the neural integrator is activated for the new eye position, which then feeds back to tonic cells that activate to hold the eye in the new position. This constitutes the step signal.

There are strong interconnections between the frontal and parietal lobes. The frontal eye fields in the precentral sulcus are involved with volitional, visually guided, purposive saccades and the parietal lobe is involved with a shift in attention to new targets appearing in the visual field. The inferior parietal lobe is involved with planning saccades and the frontal eye fields with intentional visual exploration. Other areas involved in the saccadic system include the dorsomedial supplementary motor area, which is important in learned ocular motor behaviour, and the dorsolateral frontal cortex, which is involved in the programming of saccades and attention shifts to remembered target positions.

Fibres pass from cortical areas down through the internal capsule. Below this level, there are different pathways including basal ganglia (caudate nuclei), thalamic nuclei, superior colliculi, substantia nigra, vestibular and pontine nuclei and the pedunculopontine pathway to the cerebellum (vermis and fastigial nuclei) and to brainstem nuclei (Peirrot-Deseilligny *et al.* 1995, Groh & Sparks 1996, Deleu 1997, Enderle 2002, Girard & Berthoz 2005). There is probable decussation at the level of the third nerve nuclei in the midbrain. The superior colliculi (superficial and intermediate layers) are key structures in the generation of saccades. The intermediate layer contains neurones from visual, auditory and sensory input, which are combined to guide saccades. There is a map of saccadic movements from the intermediate layer which links to the retinotopic map of visual information from the superficial layer of the superior colliculi. The basal ganglia aid control of saccades by preventing unwanted saccades (Hikosaka & Wurtz 1989).

During a saccade, the visual threshold is elevated about 0.5 log units (suppression) so that there is no awareness of the unstable visual environment during eye movement (Koerner & Schiller 1972).

The paramedian pontine reticular formation (PPRF) contains the nucleus pontis caudalis centralis, which houses the pulse generator burst cells (Keller 1974). Inhibitory burst neurones for horizontal saccades can be found caudal to the sixth nerve nuclei in the nucleus paragigantocellularis dorsalis (Scudder *et al.* 1988). Pause neurones lie in the nucleus raphe interpositus, which is located in the midline between the rootlets of the sixth nerve nucleus (Buttner-Ennever *et al.* 1988, Horn *et al.* 1996). The horizontal integrator includes the nucleus prepositus hypoglossi (NPH), the medial vestibular nucleus and other cerebellar locations (Belknap & McCrea 1988).

The rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) contains burst neurones for vertical and cyclorotational movement (Vilis *et al.* 1989, Bhidayasiri *et al.* 2000, Buttner *et al.* 2002). Bilateral input is required for vertical gaze (Bender 1980). Each burst neurone in the riMLF projects to motoneurones supplying synergistic muscle pairs, for example superior rectus and inferior oblique (Bhidayasiri *et al.* 2000). Although each riMLF contains neurones for up- and downgaze, the nucleus contains neurones only in one direction for cyclorotational quick phases (e.g. right riMLF projects clockwise and left anticlockwise). Both riMLF nuclei are connected via the posterior commissure.

Upgaze commands pass from the riMLF dorsally through the posterior commissure to the third nerve nuclei. Downgaze commands pass dorsally and caudally to the third nerve and fourth nerve nuclei. Inhibitory neurones for vertical saccades are probably also located in the riMLF (Robinson 1964, Boghen *et al.* 1974).

The riMLF projects to the interstitial nucleus of Cajal (INC) for vertical gaze holding (neural integrator) (Bhidayasiri *et al.* 2000, Buttner *et al.* 2002). The INC also receives input from vestibular nuclei, the cerebellum (particularly the dorsal vermis) and the contralateral INC (King *et al.* 1981). Fibres pass from the vestibular nuclei for vertical eye movements to encode eye position signal and head velocity signal. These fibres are position-vestibular-pause cells (PVP). Medial vestibular nuclei and NPH contribute to vertical gaze holding but only to a small extent (Cannon & Robinson 1987) (Fig. 3.1).

Smooth pursuit system

Smooth pursuits are slow eye movements under a control system capable of continuous modification of motor output in response to visual input (Chen *et al.* 2002). For slow velocities, the eye movement tracks the target accurately with velocity matching. For faster velocities, the eye may not always match the target velocity necessitating a catch-up saccade. The major stimulus for the generation of smooth pursuit movement is a fixated target that moves across the foveal and perifoveal retina. Therefore, the input signal is retinal error velocity (Yasui & Young 1975).

Movement is evoked after a latency of approximately 125 ms. Maximal sustained pursuit velocities are about 90°/s. If a target is moving at a velocity greater than

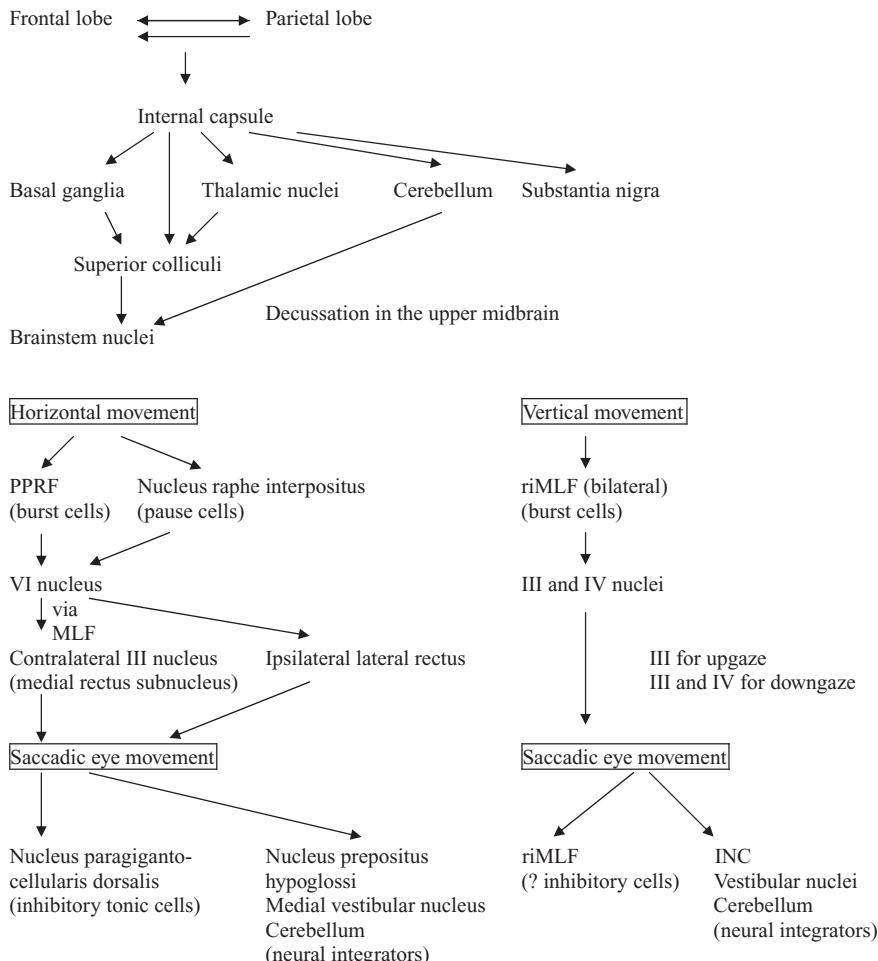


Figure 3.1 Saccadic eye movement control pathways. PPRF, paramedian pontine reticular formation; VI, sixth nerve nucleus; III, third nerve nucleus; IV, fourth nerve nucleus; riMLF, rostral interstitial nucleus of the medial longitudinal fasciculus; INC, interstitial nucleus of Cajal. (Reprinted with permission from: Rowe FJ. (2003) Supranuclear and internuclear control of eye movements. A review. *British Orthoptic Journal*, 60: 2.)

50°/s, the eyes, after lagging behind the target, may perform a saccade to catch-up before continuing with the pursuit movement. A step-ramp of innervations is the result. The step is the jump in firing frequency and the ramp is the linear increase in frequency. Latency to ramp stimulus is approximately 100 ms (Carl & Gellman 1987) and step-ramp latency is approximately 150 ms (Tychsen & Lisberger 1986).

The detailed pathways for pursuit movements are not clearly understood. The brain requires information regarding target movement relative to the position of the head. Therefore, motion processing areas of the brain are important in the

production of a smooth pursuit eye movement. Visual information is received in the visual cortex and passed to areas 17–19 (occipital and parietal cortex) including the middle temporal visual area in the superior temporal sulcus, the medial superior temporal visual area and the posterior parietal area where speed and direction of moving stimuli are encoded (Krauzlis 2004). There is a link with the frontal eye fields that contain neurones that discharge during smooth pursuit and aid programming of predictive pursuit movements, initiation and maintenance (Heinen & Liu 1997, Suzuki *et al.* 1999). Input from the basal ganglia and thalamus aid smooth pursuit control (Basso *et al.* 2005).

Efferent fibres from cortical areas pass through the internal capsule under the superior colliculi at the midbrain with a probable double decussation at the level of pons, medulla and cerebellum (Johnston *et al.* 1992), on to the dorsolateral pontine nuclei and the cerebellum (flocculus, paraflocculus and vermis) via the cerebral peduncles (Zeki *et al.* 1991). The visual information that is passed to the cerebellum is synthesised for pursuit signal and the cerebellum outputs to the vestibular nuclei that integrate the signal and passes this to the oculomotor nerves.

For horizontal smooth pursuit, the signal is directed via the dorsolateral pontine nuclei, cerebellum, nucleus reticularis tegmenti pons (NRTP) and vestibular nuclei to the sixth nerve nucleus (Suzuki *et al.* 1999). For vertical smooth pursuit, the signal is directed to the third and fourth nuclei. Medial vestibular nuclei and NPH link for velocity and position integration of horizontal smooth pursuit eye movements. The INC encodes neural integration predominantly for vertical and cyclorotatory smooth pursuit (Fig. 3.2).

Vergence system

Vergence movements are disjunctive and smooth where one eye can move independently of the other. There is continuous control over the movement generated. Vergence movements occur as a synkinesis with accommodation of the lens and pupillary constriction. The medial and lateral recti motoneurones are reciprocally innervated during vergence movements.

The stimuli for this system include disparity between the location of images on the retina of each eye, which results in fusional vergence and retinal blur caused by a loss of focus of images, which results in accommodative vergence. There is target displacement or motion along the z-axis (to or from the observer). There is generally a latency of 160 ms and maximum velocity of 20°/s in the generation of vergence eye movements.

Higher control areas for the generation of vergence eye movements are poorly understood. Vergence eye movements may occur as saccadic or smooth pursuit movements and therefore cortical areas relating to the generation of these eye movement systems will be involved in the cortical processing of visual information necessitating a vergence eye movement response.

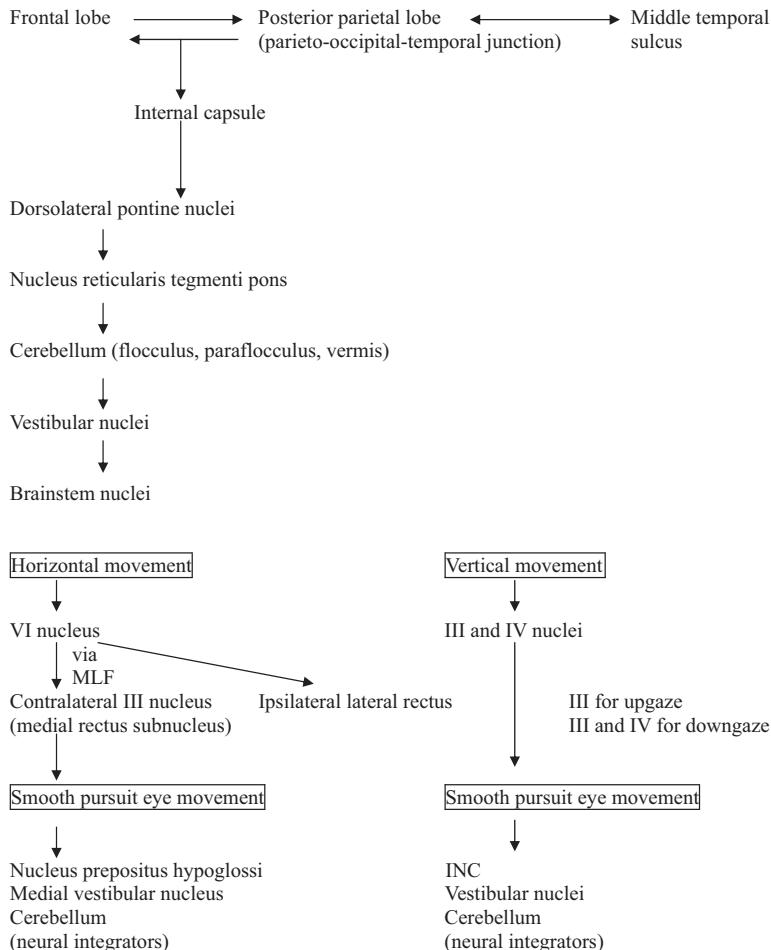


Figure 3.2 Smooth pursuit eye movement control pathways. VI, sixth nerve nucleus; III, third nerve nucleus; IV, fourth nerve nucleus; INC, interstitial nucleus of Cajal. (Reprinted with permission from: Rowe FJ. (2003) Supranuclear and internuclear control of eye movements. A review. *British Orthoptic Journal*, 60: 2.)

Neurones for control of vergence have been found in mesencephalic reticular formation (Judge & Cumming 1986), which is dorsolateral to the third nerve nuclei and contain different types of neurones; vergence tonic cells discharge in relation to vergence angle, vergence burst cells discharge in relation to vergence velocity, vergence burst-tonic cells discharge in relation to both vergence angle and velocity. These cells may serve as a vergence integrator also (Mays *et al.* 1986). Lesions near the midbrain-diencephalic junction have been documented in pseudoabducens palsy, which has been proposed as a manifestation of abnormal vergence activity (Pullicino *et al.* 2000). They postulated that inhibitory descending pathways for convergence may pass through the thalamus and decussate in the subthalamic region.

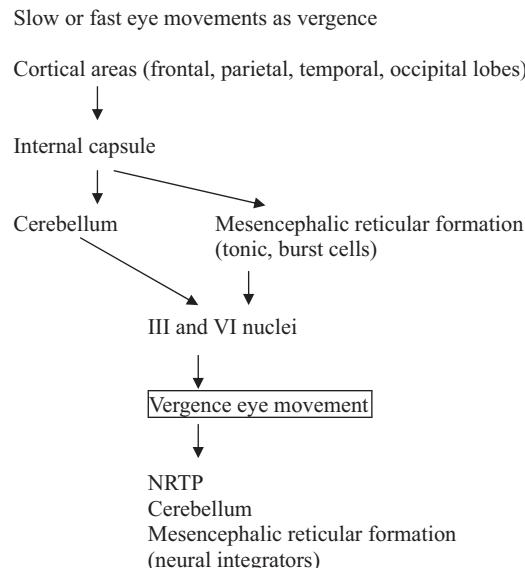


Figure 3.3 Vergence eye-movement-control pathways. III, Third nerve nucleus; IV, fourth nerve nucleus; NRTP, nucleus reticularis tegmenti pons. (Reprinted with permission from: Rowe FJ. (2003) Supranuclear and internuclear control of eye movements. A review. *British Orthoptic Journal*, 60: 2.)

Medial recti nuclei and sixth nuclei discharge for vergence eye movements (Mays *et al.* 1984). Medial rectus motoneurones are organised into different groups (A, B, and C). Subgroup C may be specifically involved in vergence commands (Warwick 1953, Buttner-Ennever & Akert 1981). There is also some cerebellar input to vergence control (Miles *et al.* 1980). The NRTP houses neurones important for generating vergence position signal which may therefore serve as a vergence neural integrator (Gamlin & Clarke 1995) (Fig. 3.3).

Vestibular-ocular response and optokinetic response

Head movement is the stimulus for vestibulo-ocular response (VOR). The VOR generates compensatory eye movements to stabilise visual images on the retina during head movements (Rambold *et al.* 2002). The latency between the onset of sudden head movements and the resultant eye movement can be as little as 15 ms (Gauthier & Vercher 1990).

This system integrates eye and body movements and is not dependent on ocular stimulation but is concerned with head position and balance. Input is received from the following:

- *Semicircular canals*: Each set of semicircular canals influences a particular pair of eye muscles. The horizontal canals produce lateral movement,

posterior canals produce vertical movement and anterior canals produce rotary movements.

- *Neck proprioceptors.*

The head movement is synthesised by semicircular canals to produce a neural signal proportional to head velocity. Stimulation of each set of semicircular canals influences a particular pair of eye muscles; horizontal/lateral canals produce lateral movement, posterior canals produce vertical movement and anterior canals produce rotary movement. The neural signal is transferred to the brainstem via the vestibular nerves and conversion of velocity data to position signal takes place in the vestibular nuclei and/or NPH. The utricle and saccule are the otolith organs. The otoliths are sensitive to tilts of the head and give rise to cyclorotational movement of the eyes to stabilise the visual image. This information is also transferred to the brainstem via the vestibular nerves to the vestibular nuclear complex where this information plus input from the cerebellum and sensory input are combined. Communication of fibres from both vestibular complexes allows continuous exchange of output for eye and head movement control.

If the head continues to rotate for more than a couple of seconds, the forces within the semicircular canals reduce to zero and no longer give a signal. The optokinetic response, which is visually driven, supplements the VOR to continue the eye movements. The optokinetic response continues indefinitely in the presence of a moving visual field to generate compensatory eye movements. There is generally a latency of 100 ms and eye velocity slowly builds to a value equal to that of the surround. The vestibulo-ocular and optokinetic responses are most often combined as the visual vestibulo-ocular reflex. There is subcortical and cortical input to optokinetic control. Retinal fibres project to the nuclei of the accessory optic tract and the nucleus of the optic tract in the pretectal area of the midbrain with subsequent projections to the pontine nuclei, NRTP, inferior olive and vestibular nuclei (Fig. 3.4).

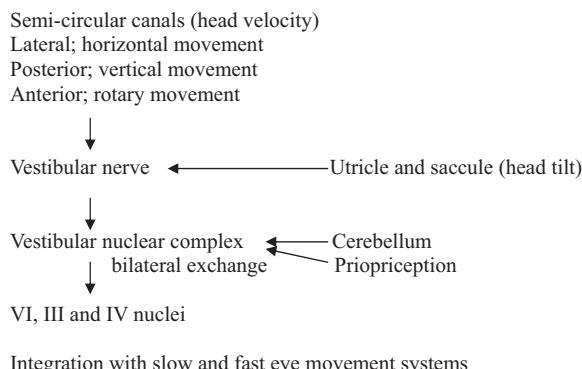


Figure 3.4 Vestibulo-ocular and optokinetic response control pathways. VI, sixth nerve nucleus; III, third nerve nucleus; IV, fourth nerve nucleus. (Reprinted with permission from: Rowe FJ. (2003) Supranuclear and internuclear control of eye movements. A review. *British Orthoptic Journal*, 60: 2.)

Brainstem control

Figures 3.5 and 3.6 depict the location of brainstem nuclei involved in the generation of eye movements. Figure 3.7 depicts cortical areas involved in the generation of saccadic, smooth pursuit and vergence eye movements and which project to brainstem nuclei.

Horizontal gaze movements

The sixth nuclei control horizontal conjugate gaze (Highstein & Baker 1978, Pola & Robinson 1978). The sixth nuclei govern conjugate movement of both the ipsilateral lateral rectus and contralateral medial rectus. The nuclei house two neurones; abducens motoneurones to the lateral rectus, and abducens internuclear neurones via the MLF to the contralateral medial rectus motoneurones in the third nuclei (Highstein & Baker 1978, Strassman *et al.* 1986).

For horizontal saccadic eye movement, saccadic commands originate from burst neurones of the PPRF, which then project to the sixth nerve nuclei. A descending smooth pursuit pathway probably projects to the sixth nucleus via the flocculus and

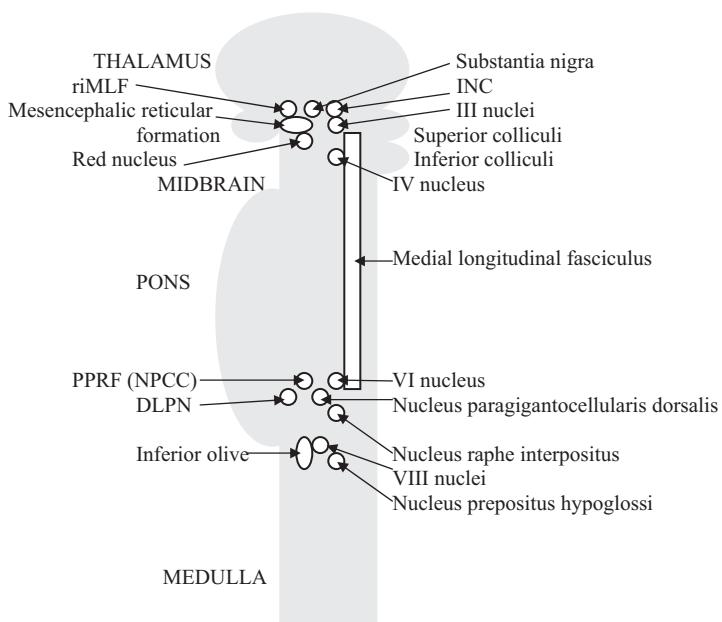


Figure 3.5 Sagittal cross section of brainstem; schematic representation. riMLF, rostral interstitial nucleus of the medial longitudinal fasciculus; PPRF, paramedian pontine reticular formation; NPCC, nucleus pontis caudalis centralis; DLPN, dorsolateral pontine nuclei; INC, interstitial nucleus of Cajal; VI, sixth nerve nucleus; III, third nerve nucleus; IV, fourth nerve nucleus; VIII, vestibular nucleus. (Adapted from Rowe FJ. (2003) Supranuclear and internuclear control of eye movements. A review. *British Orthoptic Journal*, 60: 2.)

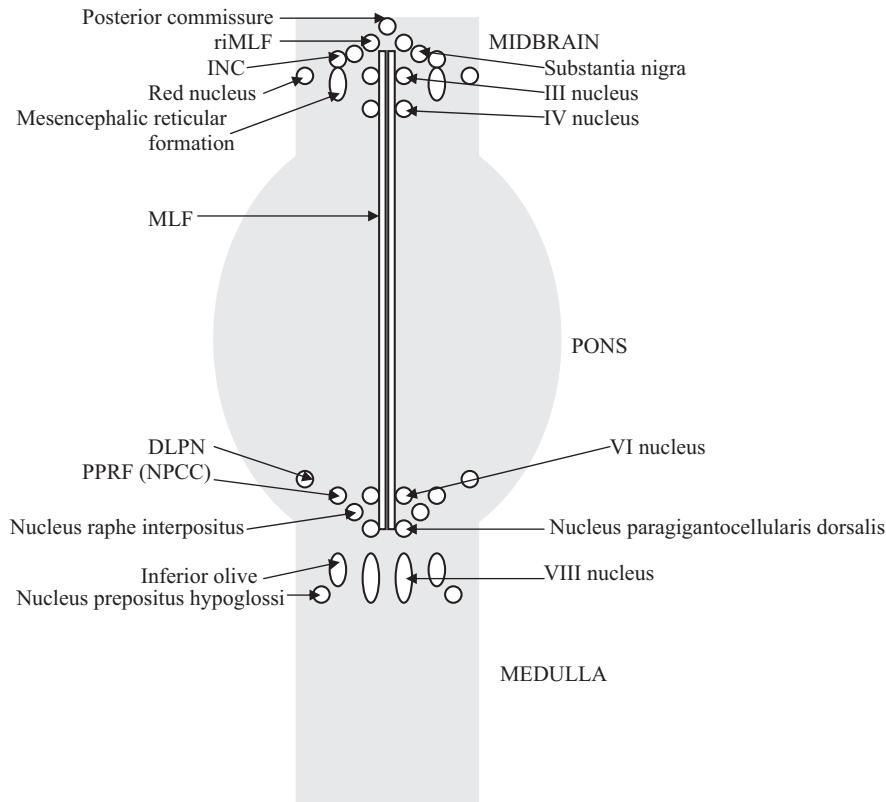


Figure 3.6 Coronal cross section of brainstem; schematic representation. riMLF, rostral interstitial nucleus of the medial longitudinal fasciculus; INC, interstitial nucleus of Cajal; MLF, medial longitudinal fasciculus; DLPN, dorsolateral pontine nuclei; PPRF, paramedian pontine reticular formation; NPCC, nucleus pontis caudalis centralis; VI, sixth nerve nucleus; III, third nerve nucleus; IV, fourth nerve nucleus; VIII, vestibular nucleus. (Adapted from Rowe FJ. (2003) Supranuclear and internuclear control of eye movements. A review. *British Orthoptic Journal*, 60: 2.)

cerebellar fastigial nuclei (Leigh & Zee 1991). The sixth nuclei receive vestibular and optokinetic inputs from the vestibular nuclei (McCrea *et al.* 1987).

Vertical and cyclorotatory gaze movements

Vertical gaze is under bilateral control from the cerebral cortex. Ocular motoneurones are in the third and fourth nerve nuclei. For vertical saccadic eye movement, saccadic generation is from riMLF (Moschovakis *et al.* 1991). Neural signals for vertical vestibular and smooth pursuit eye movements ascend from the cerebellum, medulla and pons to midbrain oculo motor nuclei via the medial longitudinal fasciculus but also the superior cerebellar peduncle.

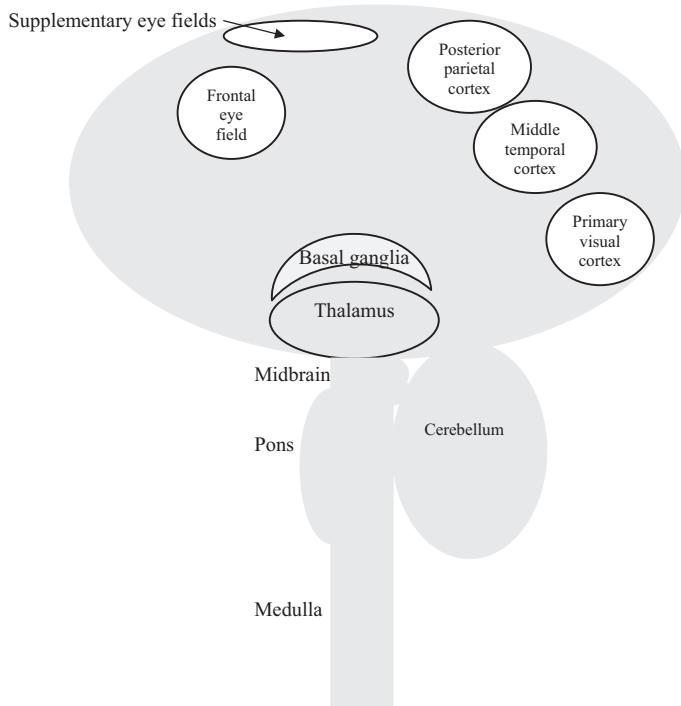


Figure 3.7 Sagittal view of cortical areas; schematic representation. (Adapted from Rowe FJ. (2003) Supranuclear and internuclear control of eye movements. A review. *British Orthoptic Journal*, 60: 2.)

Vergence gaze movements

Premotor signals for vergence are found in neurones in the mesencephalic reticular formation near the third nerve nuclei. The NRTP is important for generating vergence position signal as the vergence integrator. Sixth and third neurones coordinate conjugate and vergence commands.

Muscle sequelae

There are two relevant laws when assessing muscle sequelae:

- *Hering's law of equal innervation:* When a nervous impulse is sent to an ocular muscle to contract, an equal impulse is sent to its contralateral synergist to contract also.
- *Sherrington's law of reciprocal innervation:* When a muscle receives a nervous impulse to contract, an equal impulse is received by its antagonist to relax.

Every contraction of a muscle brings about a movement. The primary moving muscle is called an *agonist*. A movement in the opposite direction to that produced

by an agonist is caused by its antagonist. Two muscles moving an eye in the same direction are synergists.

The *primary angle of deviation* is the deviation observed when fixing with the unaffected eye in paralytic incomitant strabismus. The *secondary angle of deviation* is the deviation observed when fixing with the affected eye in paralytic incomitant strabismus.

In the course of a paresis/paralysis, Hering and Sherrington's laws cause changes in the actions of other extraocular muscles. The extent to which this develops in different patients may vary markedly. The speed and amount of development can depend partly on which eye the patient prefers to use for fixation. Generally this is the normal eye, but in some cases the paretic eye is preferred.

There are three stages in the development of muscle sequelae:

1. *Overaction of the contralateral synergist*: According to Hering's law, both the paretic muscle and its yoke (synergist) muscle will receive an equal amount of innervation. Therefore, due to the paresis, the affected eye will require extra innervation to fixate and this innervation will also be received by the synergist resulting in overaction of this muscle.
2. *Overaction of the ipsilateral antagonist*: The antagonist of the affected muscle is acting more/less unopposed, depending on the extent of the paresis, and therefore it overacts.
3. *Secondary underaction of the contralateral antagonist*: This occurs for two reasons. Since the direct antagonist of the affected muscle is acting more/less unopposed, this muscle requires less than normal innervation to move into its field of action. According to Hering's law, its yoke muscle will also receive the same little innervation and therefore, underacts. The contralateral synergist of the affected muscle is overacting. According to Sherrington's law, its direct antagonist will receive an equal impulse to relax and since it is a normal muscle, this results in underaction of the muscle.

For example, in right IV nerve palsy:

primary underaction = right superior oblique

overaction of contralateral synergist = left inferior rectus

overaction of ipsilateral antagonist = right inferior oblique

secondary underaction of contralateral antagonist = left superior rectus

Past-pointing

This is the false localisation of objects in space, in patients with recent paresis, due to the sensory changes resulting from the altered alignment of the eyes. If the patient is asked to point at an object in the field of action of the paretic muscle while the sound eye is covered, he will point beyond the object (Ambrose & von Noorden 1976).

Bell's phenomenon

In most individuals, the eyes normally rotate up and out when both lids are closed. This is demonstrated by asking the patient to try to close the lids while the examiner holds the lids open or by asking the patient to keep the lids closed while the examiner tries to open them. Presence of Bell's phenomenon indicates an intact infranuclear pathway and that the elevating muscles are receiving innervation.

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4

Orthoptic Investigative Procedures

This chapter deals with the methods used to detect the presence of strabismus or abnormal ocular movements, to measure any deviation and to assess binocular function. Methods of assessing and measuring visual acuity and additional tests for investigation of incomitant strabismus are also discussed.

Visual acuity

The visual perception of an object is resolved according to light sense, form sense and colour sense. Light sense is the absolute threshold of light. Form sense may be divided into central (the ability to discriminate fine high contrast detail) and peripheral (the field of vision) vision. Form sense is inclusive of visual acuity and contrast sensitivity.

Central visual acuity is dependent on the following:

- *The minimal visible:* This is based on the target subtending the smallest angle, which elicits a positive visual response.
- *The minimal separable:* This requires the identification of two sharp, black edges separated by a white interval of known dimension.

Visual acuity is a function of the dioptric apparatus of the eye and also of the retina, the nervous pathways and central nervous mechanisms. It is determined by the smallest retinal image whose form can be appreciated. This is the minimal separable angle with a threshold of 2–10 seconds of arc ('") and is measured by the smallest object that can be clearly seen at a certain distance. The minimal visible angle is produced at the nodal point with a threshold of 1" and represents the diameter of a cone, which is approximately 0.004 mm.

For two separate points to be distinguished, it is necessary for their images to be formed upon cones that are not adjacent to one another, but are separated by at least one cone that is unstimulated. In order to produce this, the object of fixation must subtend a visual angle of 1 minute of arc (') at the nodal point of the eye.

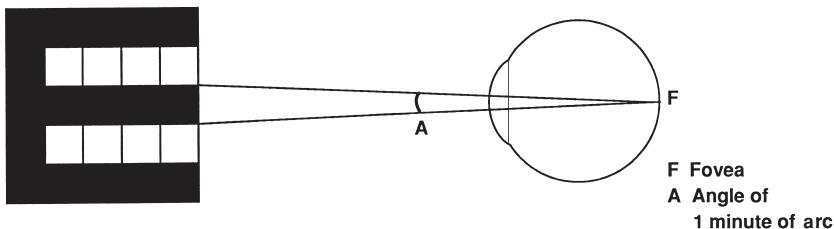


Figure 4.1 Optics of visual acuity.

This is taken as the minimum visual angle of the normal eye or minimal resolvable angle. Spatial resolution is the ability to identify that two objects are separated in space (Fig. 4.1).

Variables affecting visual acuity include:

- retinal stimulation (central stimulation involves the fovea with higher acuity function);
- luminance (brightness of target);
- contrast (sensitivity to background lighting);
- eye movements (even when looking steadily at targets there are constant refixation movements to maintain central fixation);
- contour interaction (it is easier to identify a target presented singly than one with other surrounding stimuli);
- occlusion (this may be by hand, atropine, high plus lens, blenderm, opaque glasses, opaque patch, opaque occlude or Spielmann occluder).

Clinical vision tests

Qualitative (observation of visual response)

Fixation: Observe pursuit movement and fixation preference (Calcutt 1995). Fixation preference testing reliability detects amblyopia only if the deviation exceeds 10 prism dioptres (Zipf 1976) but there is poor correlation with visual acuity. Fixation preference in the presence of strabismus may show equal alternation, fixation held through a blink, fixation held briefly, fixation held with difficulty or no fixation held with the strabismic eye.

Cover test: Unioocular fixation and objection to occlusion.

Visually directed reaching: Reaction to visual stimulus by reaching towards the object of interest.

100s and 1000s: Response to small sweets and indicates acuity of approximately 6/24.

Catford drum (Atkinson et al. 1981): Moving dots or gratings.

Stycar rolling/mounted balls: Graded balls in various sizes either mounted or rolled along the floor.

Stycar matching toys.

Ten dioptre prism (Wright *et al.* 1981): The 10 prism dioptre test is specifically designed to assess fixation preference in preverbal children who are not strabismic or who have small deviations. It may be performed base down or base up. It is recommended that the prism be held base up in cases of ptosis to aid accuracy in detecting eye movement response (Whittaker *et al.* 2000). A 25-dioptre base-in prism may also be used (Cassin 1992).

Mirror test: The infant is held close to a mirror so that they can look at their own reflection. The distance of the infant from the mirror is increased until they no longer look at themselves. Such responses have been compared to Teller acuity cards and a linear correlation is shown (Bowman *et al.* 2010). The test is useful as it is portable and easy to administer.

Quantitative (measurement of visual response)

Forced choice preferential looking cards (FCPL) (Katz & Sireteanu 1990) (Fig. 4.2): The principle of this test is that a child prefers to look at a patterned background than at a plain background (Fantz 1958, Dobson & Teller 1978, Quinn *et al.* 1993). The test cards consist of a black and white grating placed on the left or right of the card. The child's response to look towards the pattern is noted. The gratings are graded in size. FCPL may be used with a screen to exclude observer bias. The observer sits behind the screen and the baby in front. The observer records the direction of head movements in response to the appearance of the striped stimulus.

Visual acuities determined with FCPL range from approximately 6/240 in the newborn to 6/60 at 3 months and 6/6 at 36 months (Mayer *et al.* 1995). Grating acuity is better than recognition acuity (Mayer *et al.* 1984) and this difference is exaggerated in children with amblyopia (Mayer 1986, Ellis *et al.* 1988, Katz & Sireteanu 1990, Sireteanu *et al.* 1990).

Teller acuity cards (Fig. 4.3): Teller (1997) introduced a set of 16 cards that consisted of a grey background with a square wave grating on one side. An FCPL process is undertaken at a testing distance of 38 or 55 cm. Eye movements are observed towards the position of the stripes (Mayer *et al.* 1995, Salamao *et al.* 1995).

Keeler cards: Moseley *et al.* (1988) introduced a circular pattern of square wave gratings to prevent identification of the pattern by its edge. A white circle is

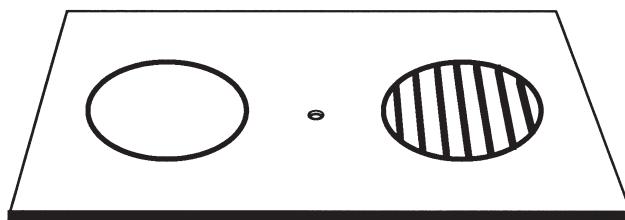


Figure 4.2 Forced choice preferential looking.



Figure 4.3 Teller cards.

present on the opposite side to the square wave pattern and an FCPL process is undertaken.

Lea gratings: Four paddles are available in this assessment with gratings printed on each side of the paddles of 0.25, 0.5, 1.0, 2.0, 4.0 and 8.0 cycles per cm. An FCPL process is undertaken to discriminate the level of grating seen.

LogMAR chart; LOGarithm of the Minimal Angle of Resolution (Sloan 1959, Frank 1986) (Fig. 4.4): The Bailey-Lovie letter chart consists of letters of almost

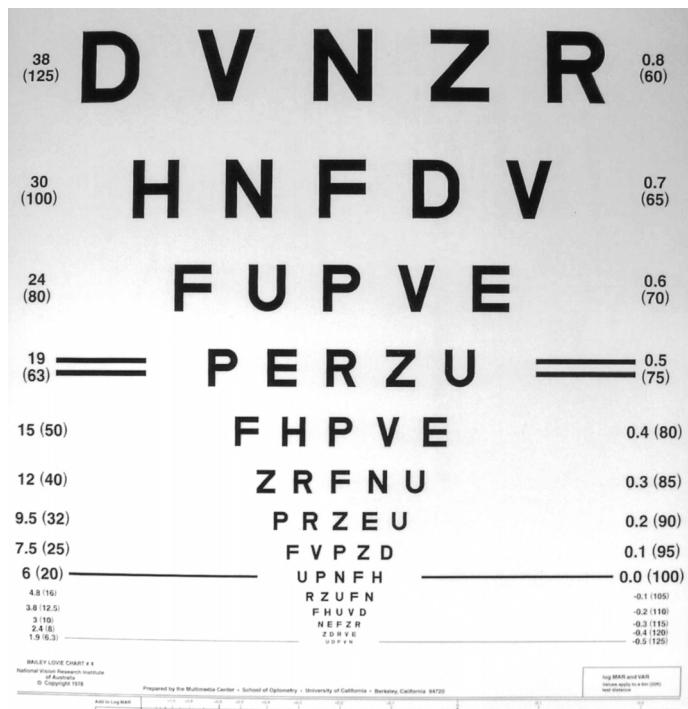


Figure 4.4 LogMAR test.

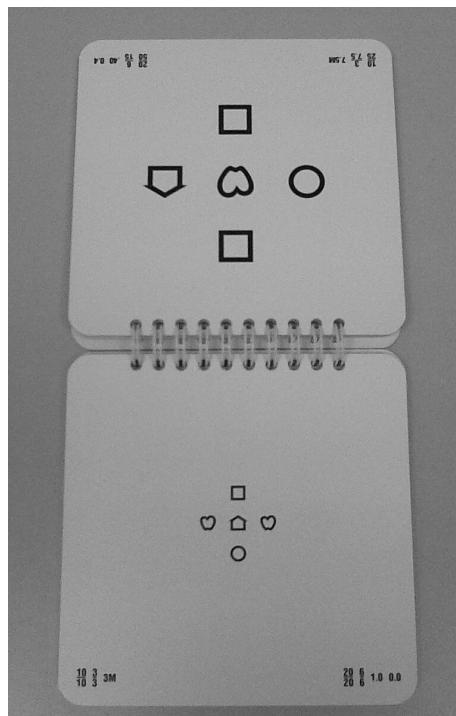


Figure 4.5 LEA symbols.

equal legibility with the same number of letters on each row. The between letter spacing is equal to one letter width and the between row spacing is equal to the height of the letters in the smaller row. The chart is designed for a standard testing distance of 6 m giving a visual acuity range of 6/60 to 6/3, the log unit for 6/60 acuity being 1.0 and for 6/6, 0. Each letter equates to 0.02 logMAR. This test is considered one of the more accurate forms of visual acuity assessment.

Further logMAR charts include ETDRS (Early Treatment Diabetic Retinopathy Study), which is useful for assessment of older children and adults (Elliott *et al.* 1995, Manny *et al.* 2003), crowded and uncrowded Keeler logMAR tests (Stewart *et al.* 2000, Jones *et al.* 2003, Shea & Gaccon 2006, Stewart 2006), crowded logMAR Kay picture test (Elliott & Firth 2009) and LEA symbols (Fig. 4.5: Becker 2002, Dobson 2003, Ruttum & Dahlgren 2006). Table 4.1 provides information on age-related normal visual acuity results.

Snellen test type (Fig. 4.6): This test is the most widespread used assessment of visual acuity. It consists of a chart of letters starting with one letter at the 6/60 level and with additional letters on each line progressing to the 6/4 level. There is no related letter spacing or row spacing according to the size of letters on each line, which renders the test less accurate than logMAR (Stewart 2006).

Sheridan Gardiner singles/linear test type (Fig. 4.7): This test uses seven letters (X, U, T, O, H, A, V). The letters are presented singly on a card or as a linear chart in

Table 4.1 Age-related visual acuity norms.

Chart	Age range	Visual acuity	Reference
Crowded Keeler logMAR	3 years	0.2 ± 0.09 Range: 0.025–0.375	Shea <i>et al.</i> 2006
Crowded Keeler logMAR	4 years	0.142 ± 0.08 Range: −0.025–0.3	Shea <i>et al.</i> 2006
Crowded Keeler logMAR	3–5 years	0.04 Range: −0.125–0.3	Jones <i>et al.</i> 2003
Crowded Keeler logMAR	4–6 years	0.057 ± 0.10 Range: 0.0–0.4	Stewart 2000
Uncrowded single Keeler	4–6 years	−0.01 ± 0.10 Range: −0.1–0.3	Stewart 2000
ETDRS	6–11 years	0.01 ± 0.08 Range: −0.16–0.14	Manny <i>et al.</i> 2003
ETDRS	17–18 years	−0.1	Ohlsson 2003
ETDRS	18–24 years	−0.13 ± 0.06 Range: −0.2–0.0	Elliott <i>et al.</i> 1995
Kay linear log	3–5 years	−0.04 Range: −0.1–0.225	Jones <i>et al.</i> 2003
Crowded Kay Picture test	3.5–4 years	0.05 Range: 0.0–0.10	Cross <i>et al.</i> 2010

**Figure 4.6** Snellen test.



Figure 4.7 Sheridan Gardiner test.

rows, and the patient is required to match the letters with those on a key card. The letters are graded from 6/60 to 6/3.

Kay's pictures (Fig. 4.8): A variety of pictures that are graded in size are presented singly on a card. The patient is required to identify the picture by naming it or matching it with a key card. These pictures range from 6/60 to 6/3. Kay pictures are also available in crowded logMAR format (Cross *et al.* 2010).



Figure 4.8 Kay's pictures.

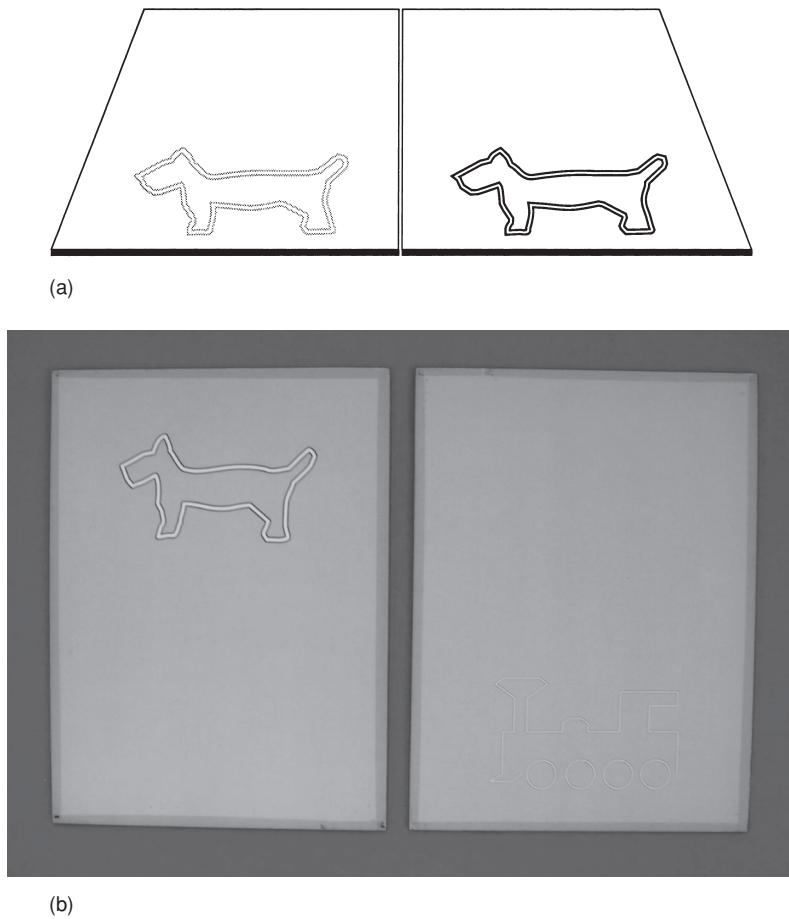


Figure 4.9 Cardiff acuity cards.

Cardiff acuity cards (Woodhouse *et al.* 1992, Horwood 1994) (Fig. 4.9): This test is based on the principle of vanishing optotypes. It consists of a series of cards displaying a picture on each. The defining outer lines that form the picture decrease in size until the picture appears to fade into the grey background. The patient's ability to recognise each picture is noted. Three card choices are available at each acuity level and the test may be held at 50 or 100 cm test distance.

Cambridge crowding cards (Atkinson *et al.* 1988): Five letters (H, O, T, V, X) are used in this test. Each letter to be matched is surrounded by the other letters, which introduces a crowding effect. The patient identifies the centre letter on a key card.

Sonksen-Silver test: This test presents six letters (X, O, U, H, V, T) in linear format, which the patient matches with letters of a key card.

Stycar letters: Three groups of letters may be presented, depending on the patient's age, and the patient is required to match the letters on a key card. The first group contains five letters (V, T, O, H, X), the second group contains seven letters (the

first five plus A and U) and the third group has nine letters (the existing seven plus L and C). The letters are graded from 6/60 to 6/4.

Ffookes symbols: This test presents the shapes of a circle, square and triangle, which the patient matches with a hand-held corresponding shape.

Illiterate E test: Capital letter E's are presented with the letter directed up, down, right or left. The patient identifies the relevant orientation of the letter.

Landolt C test: This test is similar to the illiterate E test in that the capital letter C is presented with the letter directed up, down, right or left. The patient is required to indicate the position of the gap in the letter.

Sjogren hand test: A black hand with the fingers pointing up, down, right or left is presented and the patient identifies the orientation of the position of the fingers.

Near tests

1. Reduced Snellen test
2. Reduced Sheridan Gardiner
3. Reduced E test
4. Maclare book
5. Moorfields bar reading book
6. N series test

Pinhole test

This can be used to assess whether or not reduced vision is caused by an uncorrected refractive error. One eye is occluded, a pinhole is placed before the other eye and the visual acuity is tested. When vision improves, this is strongly indicative of a refractive error. If no improvement occurs, there is amblyopia or some organic cause for the reduced vision.

Crowding phenomenon

It is easier to see a single optotype than a letter on a linear chart. The reduced visual acuity is due to the additional stimuli from surrounding letters and results in confusion. Crowding is specifically caused by contour interaction and attentional factors (Leat *et al.* 1999). This is noted in particular with reduced visual acuity in amblyopes (see Chapter 8).

Age indications for use of visual acuity tests

Age	Test
0–6/12	Forced choice preferential looking, visual evoked response, optokinetic nystagmus, observation, fixation, pursuit, visually directed reaching, objection to occlusion, cover test, Catford drum.

Age	Test
6/12–2 years	100s and 1000s, visually directed reaching, Stycar balls, forced choice preferential looking, Cardiff acuity cards.
2–3 years	Kay's pictures, Stycar toys, illiterate E test, ffookes symbols, Sjogrens hand test, logMAR.
3+ years	Sheridan Gardiner test, Cambridge crowding cards, Sonksten-Silver test, Landolt C test, Stycar letters, Snellen test, logMAR.

Contrast sensitivity

Contrast sensitivity function is the ability to distinguish between different levels of contrast. It is a measure of the limit of visibility for low contrast patterns. Contrast sensitivity uses sine waves, which are continuous waveforms that oscillate in a smooth and regular manner and can be of different frequencies and amplitudes. Spatial frequency is the number of cycles per degree: one cycle includes one dark and one light area.

Contrast sensitivity is calculated as 1/contrast threshold; the threshold being the level at which contrast can just be seen. Detection of contrast relates to the organisation of the receptive field, diffraction, aberrations, scatter, cone density, central versus eccentric viewing, luminance and presence of ocular disease.

Contrast sensitivity may be more related to visual performance and more sensitive than visual acuity as an assessment of visual function. Reduced contrast results in symptoms even if there is a normal level of visual acuity. Similarly, improvements in contrast sensitivity can occur despite no obvious change in visual acuity.

Contrast sensitivity tests

Pelli-Robson chart: This test uses letters of low spatial frequency. There are eight lines with groups of 3 letters. Contrast reduces between each grouping. The test records maximum contrast sensitivity (Elliott *et al.* 1990).

VISTECH (Fig. 4.10): This chart contains 40 circles of different orientations. Contrast reduces from left to right along five rows of differing spatial frequencies (Rogers *et al.* 1987).

Cambridge test: Twelve plates contain spatial frequencies of 4 cycles/degree. The patient makes a forced choice preferential looking between the plate and a blank.

Arden gratings: Seven plates contain different spatial frequencies and with reducing contrast. Each plate had a single spatial frequency with the grating oriented vertically. Contrast varies from high to low and from bottom to top.

Melbourne edge test: Twenty plates contain reducing contrast. The patient identifies the orientation of the edge.

Computer-generated contrast: Different spatial frequencies and contrasts are presented on a computer screen. The patient indicates when the grating is visible.

Lea contrast (Fig. 4.11): The patient reads numbers in decreasing contrast until unable to see any more.

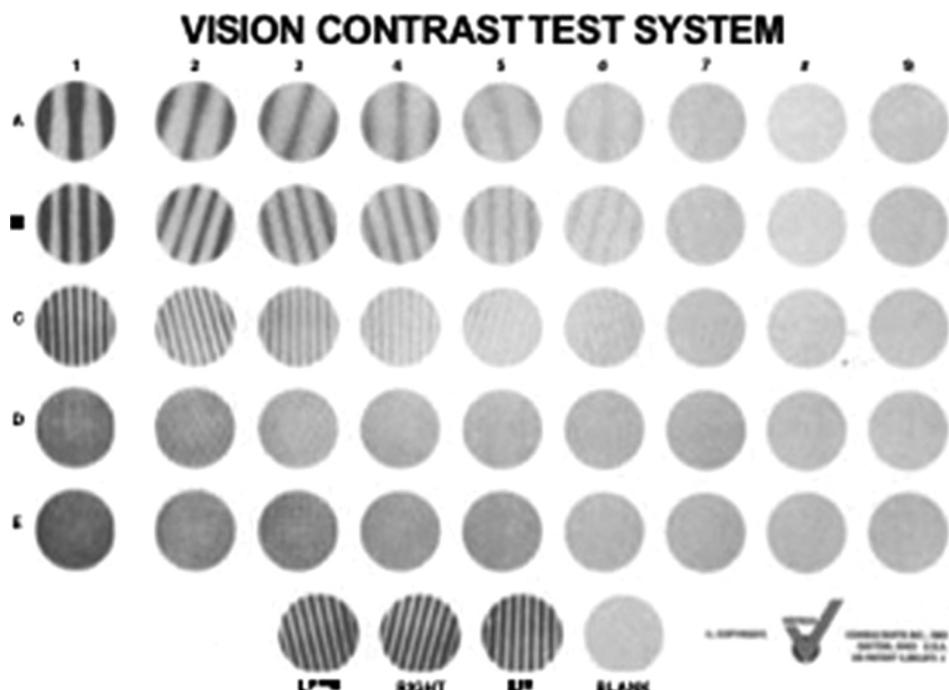


Figure 4.10 Vistech chart.

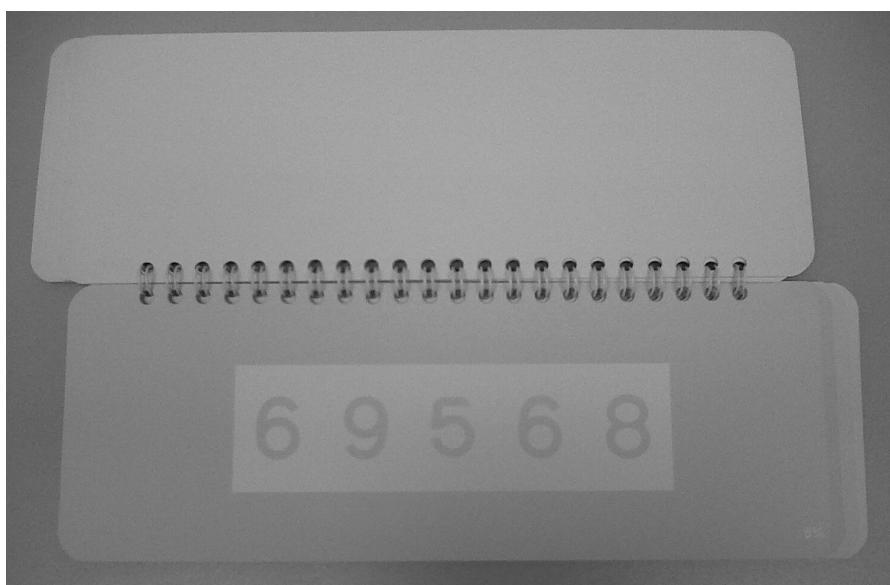


Figure 4.11 LEA contrast numbers.

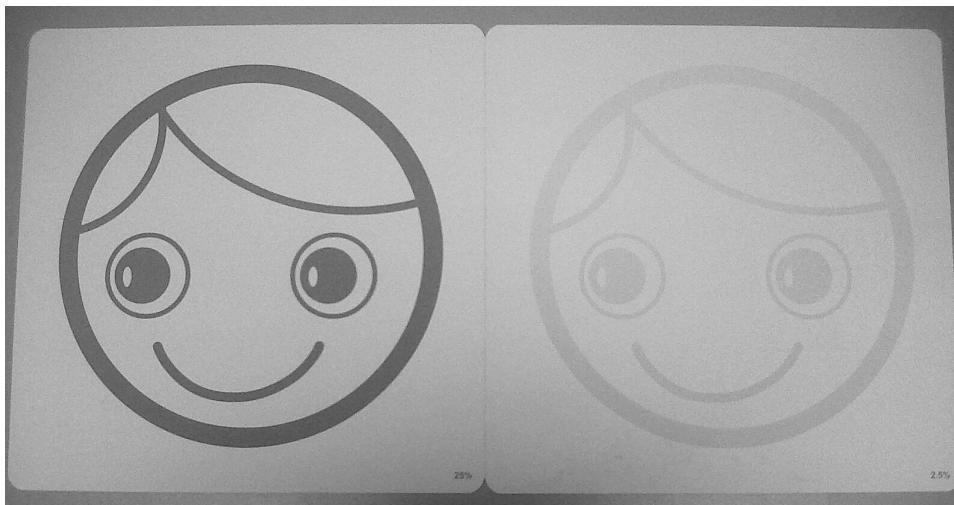


Figure 4.12 Heidi contrast faces.

Hiding Heidi (Fig. 4.12): The child must detect when the face disappears over a range of different contrasts. There are four cards with a face printed on either side on differing contrast levels from black, 25%, 10%, 5%, 2.5% to 1.25%.

Refractive errors

Assessment of the presence of refractive errors is important to the investigation and management of strabismus and amblyopia.

A cycloplegic refraction is required for infants and young children to ensure non-accommodation in order to obtain an accurate result. Usually 1% cyclopentolate is used. However, since 1% cyclopentolate may cause a transient increase in blood pressure in infants, 0.5% is used and a drop of 2.5% phenylephrine hydrochloride may be added if mydriasis is unsatisfactory. Maximum cycloplegia is obtained in one hour but can last 24 hours. Atropine 0.5 or 1% ointment or drops may also be instilled where indicated for dark, heavily pigmented irides. The effect on the ciliary muscle lasts up to 10 days, and for pupil size, it may last up to 14 days. Cycloplegic refraction will exclude accommodation and abolish ciliary muscle tone so that the total static refraction can be established. The pupils are also dilated, which facilitates refraction as the length of the retinoscope streak is increased.

Causes of refractive error include changes in the axial length of the eye, dioptric power of the lens and dioptric power of the cornea.

Emmetropisation is the process of eye growth in which the refractive index of the ocular structures (lens and cornea) and axial length of the eye balance each other resulting in emmetropia. Thus, the image of a viewed object will focus on the retina (emmetropia).

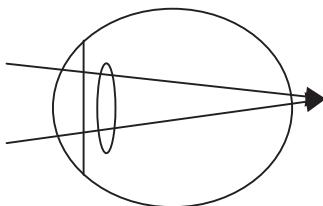


Figure 4.13 Hypermetropia.

Hypermetropia

This occurs where parallel rays of light come to a focus behind the retina (Fig. 4.13) when the eye is at rest and may be caused by a short axial length and/or insufficient refractive curvature of the lens/cornea. Total hypermetropia consists of latent and manifest hypermetropia. Latent hypermetropia can be overcome physiologically by the tone of the ciliary muscle – up to 1 dioptre. Manifest hypermetropia consists of facultative and absolute hypermetropia. Facultative can be overcome by accommodation whereas absolute cannot. A low degree of hypermetropia is considered less than 2.50 dioptres. Moderate degree is considered between 2.50 and 6.00 dioptres. High degree is considered to be greater than 6.50 dioptres. Children with greater than 3.50 dioptres of hypermetropia are up to 13 times more likely to develop strabismus (Atkinson *et al.* 2000) and have lower education scores (Williams 2005). Full correction of hypermetropia has been shown to impede (but not halt) emmetropisation compared to those not wearing glasses (Ingram *et al.* 2000). Those undercorrected by 1 dioptre show no significant difference to those not wearing glasses (Atkinson *et al.* 2000).

Myopia

This occurs where parallel rays of light come to a focus in front of the retina (Fig. 4.14) when the eye is at rest and may be caused by a long axial length and/or greater curvature of the lens/cornea. The COMET (Correction Of Myopia Evaluation Trial) study has shown that progressive addition lenses slow the progression of myopia (Gwiazda *et al.* 2003). The CLEERE (Collaborative Longitudinal Evaluation of Ethnicity and Refractive Error) study has documented that eyes of myopia patients are shaped like myopic eyes before the onset of myopia with clear ethnic

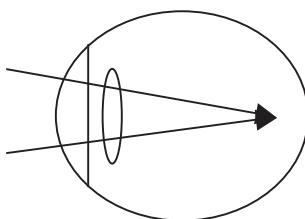


Figure 4.14 Myopia.

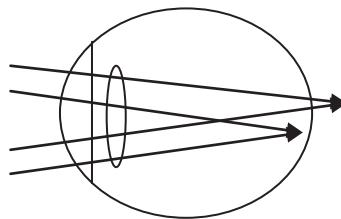


Figure 4.15 Astigmatism.

differences (Jones-Jordan *et al.* 2007). The CLAMP (Contact Lens And Myopia Progression) study has shown no long-term benefit for rigid gas permeable contact lens on progression of myopia.

Astigmatism

This refractive error occurs where a point of focus cannot be formed on the retina owing to unequal refraction in different meridians (Fig. 4.15). There is irregular curvature of the lens/cornea. A difference of greater than 1 dioptre cylinder may be considered as treatable in children over the age of 1 year.

Against the rule, astigmatism occurs when the vertical meridian is relatively hypermetropic and the horizontal meridian is relatively myopic. As a rule, astigmatism occurs where the refractive power of the vertical meridian is the greatest.

Regular astigmatism is a refractive anomaly where the two maximally differing meridians of the eye are at right angles. Oblique astigmatism has the maximally differing meridians at oblique angles. Simple astigmatism occurs when some rays focus on the retina and others are focussed behind or in front. Compound astigmatism occurs when all rays focus behind or in front of the retina. Mixed astigmatism occurs where some rays are focussed behind the retina and others in front.

Anisometropia

This is a difference in refractive error between the two eyes with a difference usually of greater than 1 dioptre.

Antimetropia

In this state, one eye is hypermetropic and the other eye is myopic.

Emmetropisation

This is the mechanism in which refractive error dissipates during early visual development of infants. Many babies are born with a small degree of hypermetropia, usually in the region of +2 dioptres. Over the following 2-year period, the hypermetropia reduces rapidly and then reduces at a slower pace over the following

6 years as the eye grows to adult size. During emmetropisation, the refractive state of the cornea and lens of the eye cancel out the axial length of the eye.

The mechanism of emmetropisation is different in children who have strabismus. These children do not have the same initial rapid reduction of hypermetropia but instead have only a slow reduction (Birch *et al.* 2010). In addition, the deviating eye has a slower reduction of hypermetropia than the fellow eye resulting in anisometropia (Ingram *et al.* 2003).

Monovision

In this refractive state, patients use one eye for distance viewing and one eye for near viewing. This may be deliberately achieved with refractive surgery. An orthoptic assessment prior to refractive surgery is essential where there is history of strabismus or accommodative deficit, amblyopia, antimetropia, previous use of prisms and aniseikonia.

Aniseikonia

A binocular condition in which the images seen by the right and left eyes differ in size or shape. It is common in patients with corrected anisometropia and also in aphakic patients, those with oblique astigmatism and pseudophakia. Each dioptre of corrected anisometropia will result in a 1% difference in size of retinal images. Individuals may cope with up to 3% difference in image size before symptoms arise. Symptoms include mild distortions of visual space, headaches, asthenopia and disrupted binocular vision (Stewart & Whittle 2002). Aniseikonia may be corrected with contact lenses, iseikonic lenses or manipulation of spectacle lenses.

Effects of ill-fitting glasses

The following may occur because of poorly fitted glasses:

- Blurred visual acuity
- Patient unwilling to wear glasses
- Disturbed muscle balance and binocular vision
- Decentration of lenses causing prismatic effect
- Tilting of spectacle frame
- Tilting of lenses
- Distance of lenses from eyes
- Twisting of lenses within frames

Modification of refractive error prescription

It is possible to alter the prescription by manipulation of the lenses in order to help with management of strabismus, accommodative or convergence deficits.

A change is produced which can induce an exertion or relaxation of accommodation. A permanent change may be obtained by altering the glasses prescription. A temporary change may be obtained by using Fresnel lenses or clip-on lenses.

Cover test

The cover test is one of the most important diagnostic tests used in orthoptic practice. This is an objective dissociative test to elicit the presence of a manifest or latent deviation. It relies on the observation of the behaviour of the eyes whilst fixation is maintained and each eye is covered and uncovered in turn.

Equipment

Occluder: A card/paddle that is opaque and large enough to cover one eye should be used so that the patient cannot see through or around it. A Spielmann occluder is a translucent occluder through which the patient can only see diffuse light without contours but with which the observer can see the patient's eyes (Spielmann 1986) (Fig. 4.16).

Fixation target: A small object that will maintain the patient's interest, such as a picture for a child and a letter for an adult should be used at 1/3 m. This will make the patient accommodate and is therefore also called an accommodative



Figure 4.16 Occluders.



Figure 4.17 Fixation targets.

target. A Snellen letter or picture at 6 m from the eyes is also required. Visual acuity must be good enough to see the fixation target with a minimum of 6/60 for Snellen's fixation. Otherwise, corneal reflections must be monitored (Fig. 4.17).

Pen torch: This should project a single spot of light.

Cover/uncover test

Manifest deviation

The test is performed both with and without spectacles and also with and without any abnormal head posture. The patient looks at a spotlight held at eye level 1/3 m away. The examiner should also be at the patient's eye level. Corneal reflections are noted to see if there is an obvious deviation.

One eye is covered with an opaque occluder and the other eye is observed. If the uncovered eye moves to take up fixation, a manifest deviation of that eye is present (Fig. 4.18). The deviation is present in the opposite direction to the take up of fixation, as in the following examples:

- Temporal movement indicates a convergent (eso-) deviation.
- Nasal movement indicates a divergent (exo-) deviation.
- Downwards movement indicates an upward (hyper-) deviation.
- Upwards movement indicates a downward (hypo-) deviation.

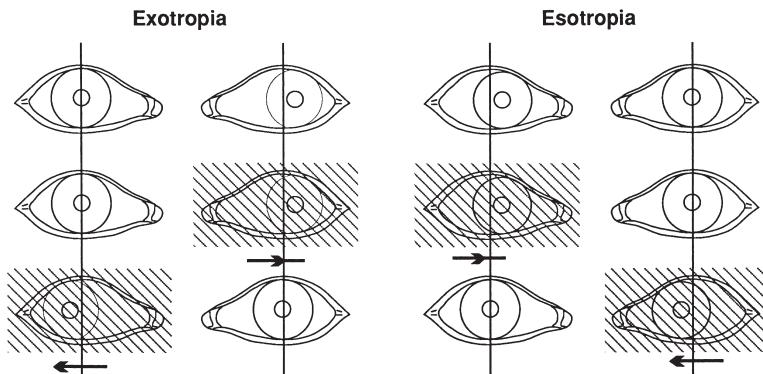


Figure 4.18 Cover/uncover test in manifest strabismus. The left exotropic eye does not move when an occluder is placed before the left eye. On occlusion of the right eye, the left eye moves inwards to take up fixation. The right esotropic eye does not move under cover of the occluder. It moves outwards to take up fixation on occlusion of the left eye.

If no movement of the uncovered eye is seen, there is usually no manifest deviation present. The exception is if eccentric fixation is present where a manifest deviation may be noted whilst observing the corneal reflections.

The test is then repeated with the other eye covered, using a fixation target placed firstly at 1/3 m and then at 6 m. A target may need to be placed at a distance of greater than 6 m if a significant increase or decrease in the angle of deviation is noted between near and distance fixation. This may occur particularly in exo-deviations. The deviation will alternate in cases of alternating heterotropia.

Latent deviation

The patient fixates a spotlight at 1/3 m. One eye is completely covered. The cover is removed and any movement of the covered eye is noted. The test is then repeated for the other eye and with fixation targets for 1/3 m and 6 m, and also for greater than 6 m where there is a significant increase or decrease in the angle of deviation between near and distance fixation, particularly in exo-deviations (Fig. 4.19).

Alternate cover test

The patient fixates a spotlight at 1/3 m. One or other eye is covered alternately throughout the test to ensure the complete dissociation, which is necessary to detect latent deviations. Movement of the uncovered eye is noted as the cover is changed from one eye to the other. The test is then repeated with a fixation target at both 1/3 m and 6 m. Finally, the cover/uncover test is repeated to ensure that full recovery and not partial recovery to a manifest deviation has occurred (Fig. 4.20).

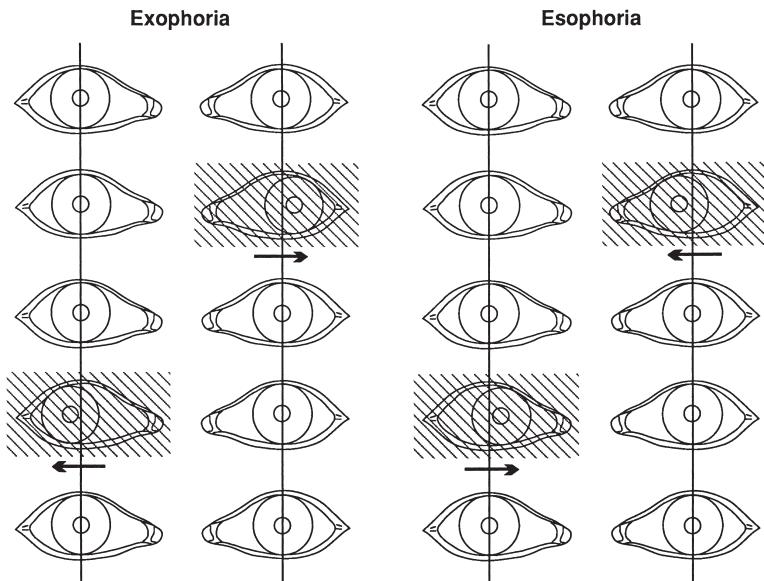


Figure 4.19 Cover/uncover test in latent strabismus. With latent deviations, the eye deviates under cover of the occluder. With exophoria, the eye moves outwards under cover and on removal of the occluder it moves inwards to refixate and binocular single vision is regained. With esophoria, the eye moves inwards under cover and on removal of the occluder it moves outwards to refixate and binocular single vision is regained.

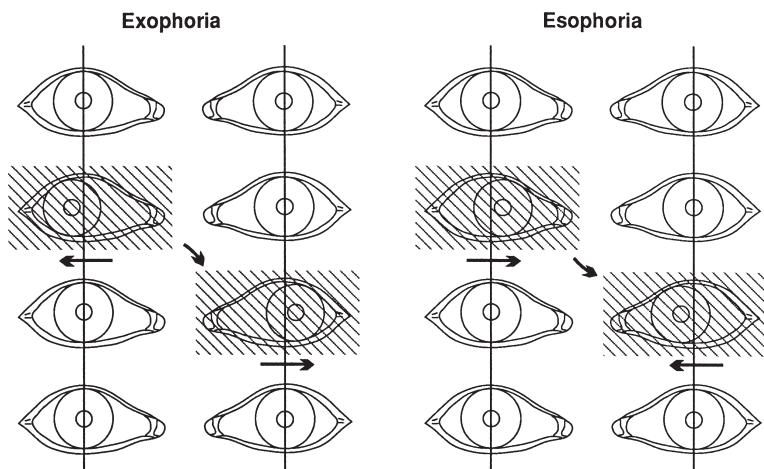


Figure 4.20 Alternate cover test. The eyes move outwards under cover with exophoria and move inwards to refixate. The eyes move inwards under cover with esophoria and move outwards to refixate.

Information gained from cover testing

Manifest deviation

1. *Type of deviation:* Eso-, exo-, hyper-, hypo- and cyclo-.
2. *Size of deviation:* Slight, small, moderate and marked.
3. *Speed to take up fixation:* This gives a good indication of the level of vision in the eye. Rapid uptake indicates the presence of reasonable vision in the deviating eye; slow uptake indicates poor vision; a wandering eye indicates non-central fixation with very poor vision. Eyes that show alternating fixation are often favoured equally and as a result have equal vision.
4. *Effect of accommodation on the deviation:* It can be determined by comparing the deviation on cover test to a light and detailed fixation target.
5. *Nystagmus:* Manifest nystagmus is seen before the cover test is performed but latent nystagmus is only elicited when one eye is covered (see Chapter 17).
6. *Dissociated vertical deviation (DVD):* See Chapter 6.
7. *Incomitance:* The angle of deviation will vary depending on which eye is covered.

Latent deviation

1. Type of deviation.
2. Size of deviation.
3. Rate of recovery to achieve binocular single vision, for example rapid, moderate, slow, delayed; deviation decompensates further to produce a manifest deviation. The rate of recovery indicates the strength of control over the deviation.
4. Partial recovery. There is recovery to a small manifest deviation – usually microtropia (see Chapter 7).

Ocular motility

Versions are rotary movements of both eyes from the primary position in the same direction. Ductions are rotary movements of one eye from the primary position.

Since the eye has an essentially fixed centre of rotation, it can rotate around one of three axes, all going through the centre of rotation (Boeder 1957) (Fig. 4.21):

- *x-axis:* Horizontal axis around which vertical movements occur, perpendicular to the line of sight.
- *y-axis:* Anterior/posterior axis for torsional movements, coincident with the line of sight.
- *z-axis:* Vertical axis for horizontal movements, perpendicular to the line of sight.

Ocular movements are poorly coordinated at birth but rapidly develop in the first 3 months of life. Range of ocular movements is largely preserved until older age

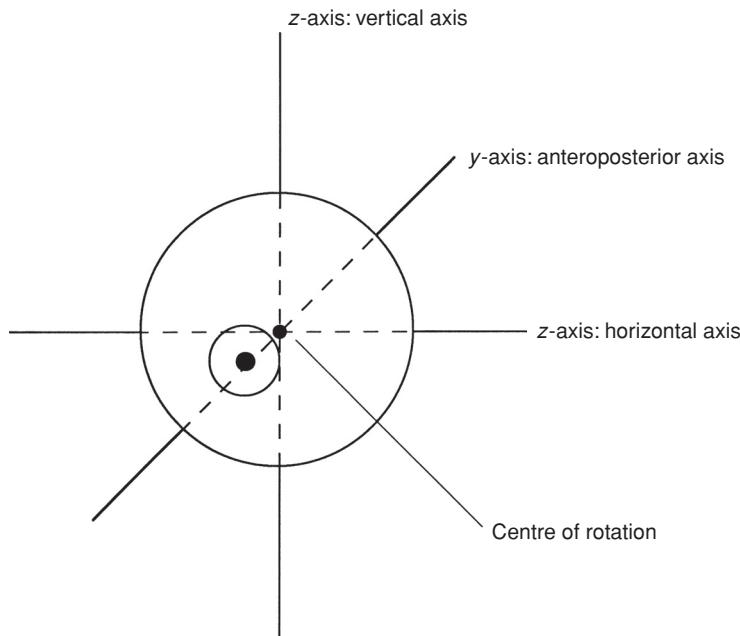


Figure 4.21 Rotation of the eye.

when there can be a reduction in elevation (Chamberlain 1970, Clark & Isenberg 2001, Davidson & Knox 2003). With ageing, there is attenuation of connective tissues around the extraocular muscle pulleys in the orbits and particularly in the orbital layer (Virdee 2007). This may result in displacement of destabilisation of the muscle pulleys in which the horizontal muscle pulleys sag inferiorly and symmetrically giving rise to limited elevation.

Saccadic movement

The patient is asked to look from one target to another, one held in the primary position and the other:

- to each side of the head for horizontal movement;
- above and below the head for vertical movement.

The targets are held a short distance apart initially (e.g. 10 cm) followed by holding them further apart (e.g. 50 cm). The movement, speed and accuracy of either eye is compared and any asymmetry between the eyes is noted.

Testing of saccadic eye movements is useful in:

- comparing movement of the two eyes as this may show slight limitations;
- suspected myasthenia gravis as fatigue may become evident;
- suspected internuclear ophthalmoplegia as dysmetria may be evident.

Smooth pursuit movement

Versions are tested binocularly using a spotlight to observe the movement from the primary position into the eight cardinal positions of gaze with the head erect and immobile. The corneal reflections are observed and a cover test is performed in each position.

Ductions are tested monocularly to compare with versions and the following noted:

- Excursion
- Behaviour of both eyes
- Whether the movement is smooth or jerky
- Nystagmus
- Change in position of the lids or globe
- Effect of fatigue
- Torsional movement
- Any discomfort on movement
- Abnormal head movements

Ocular movements may be recorded in written or graphic form (see ‘Illustrative Case Report’ Section).

Optokinetic movement

This assesses the integrity of both saccadic and smooth pursuit pathways by movement of high contrast repetitive targets (black and white) across the field of vision.

Optokinetic nystagmus (OKN) may be tested with an OKN/Barany drum (Fig. 4.22) or OKN scarf. It is tested horizontally with movement right and left and vertically with movement up and down. The patient is asked to watch the stripes in turn and the following noted:

- Movement present or absent
- Difference between either eye
- Difference in movement to either side or up and down
- Smooth pursuit movement (slow phase)
- Saccadic movement (fast phase)

Vestibular movement

Doll's head testing

This is a rapid, passive head rotation used to elicit an initial contraversion deviation of the eyes followed by a prompt recentering. The head is moved quickly by the

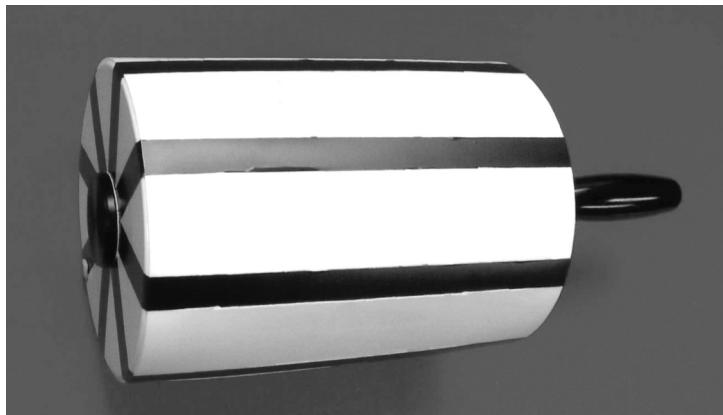


Figure 4.22 Optokinetic nystagmus drum.

examiner to the right and left for horizontal movement and up and down for vertical movement. A normal response is of the eyes moving in the opposite direction to the head movement.

Swinging baby test

A similar response to the Doll's head test is seen. It is based on the observation of a conjugate deviation of the eyes in response to head movement induced by rotation and is useful with babies who will not respond to conventional testing.

The infant is held upright facing the examiner. The examiner rotates him/herself and the infant through 360° while observing the infant's eyes. An indication of the extent of ocular movement is gained and also an indication of the level of visual acuity is gained. Post-rotational nystagmus in sighted babies will only persist for a few seconds before being suppressed.

Induced vestibular nystagmus

This results from altered input from the VIII nerve. Caloric testing alters input by instilling warm or cold water into one ear. Warm water produces a slow phase to the opposite side and fast phase to the same side. Cold water produces a slow phase to the same side and a fast phase to the opposite side. Vertical caloric nystagmus is induced by stimulating both ears. Rotational testing produces nystagmus, which is induced by seating the patient in a Barany chair with the head forward 30° and rotating the chair. By rotating the chair to the right, there is a fast phase to the right and slow phase to the left. Post-rotational nystagmus in the opposite direction follows on ceasing the rotation. The patient is aware of this movement and may feel nauseated.

Quantitative assessment of eye movements

There are numerous methods in which the eye movements of subjects can be quantitatively assessed:

Electro oculogram (EOG): The EOG is a measure of biopotentials resulting from changes in eye position. There is an electrical potential difference between the two poles of the eye: cornea and retina. Eye movement can be detected by measurement of changes in this potential by electrodes placed to either side of the eye as the eye moves. It is a cheap method of eye movement measurement, non-invasive and relatively easy to use. However, it does not offer high temporal or spatial resolution and is prone to artifacts.

Infrared oculography: This technique measures the reflection of infrared light projected onto the eye. Any asymmetry in light reflection from the two eyes can thus be quantified. It offers good spatial resolution and is not affected by ambient lighting. However, it can be an expensive system and blinks interrupt measurement.

Video-based measurement: Eye movements are measured by the digital processing of video images of the two eyes. The position of the eyes is established by detecting the pupil and iris landmarks and three-dimensional rotational angles of the eyes are established.

Scleral search coils: These are used to measure eye position. A wire coil is embedded in a contact lens or annulus and when placed on the eye, induces a signal of the eye position when in a magnetic field. The magnetic field is generated by additional coils placed on either side of the head (top and bottom, right and left for vertical and horizontal movements). Coils can be used to measure horizontal, vertical and rotator eye movements with high temporal and spatial resolution. The system is expensive and invasive.

Accommodation and convergence

Accommodation

Accommodation is the ability to increase the convexity of the crystalline lens in order to obtain a clear image of a near object. It is closely associated with convergence and pupil constriction. The *far point of accommodation* is the furthest distance at which an object can be seen clearly and the *near point*, the nearest point at which an object can be seen clearly. The *range of accommodation* is the distance over which accommodation is effective; the distance between the near and far points. The *amplitude of accommodation* is the difference in the focus power of the eye whilst fixating for near distance and fixating for far distance. Relative accommodation is the amount by which accommodation can be increased or relaxed whilst convergence remains constant.

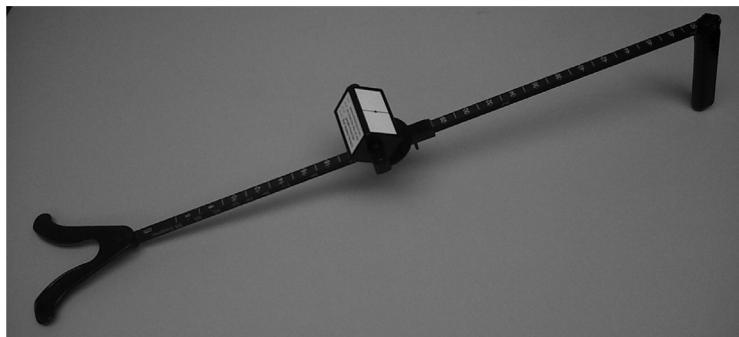


Figure 4.23 RAF rule.

Measurement of accommodation

Accommodation is measured in dioptres. One dioptre has a focal distance of 1 m and therefore accommodation is calculated in terms of the reciprocal of the fixation distance. The *near point of accommodation* is assessed using the RAF rule (Fig. 4.23). The rule cheek pads are placed on the patient's cheeks and held in a slightly depressed reading position. The patient wears distance spectacles (if any) but may need to repeat the test using reading spectacles. Using the N series target, the patient observes the smallest line seen with either eye.

With both eyes open, the carrier is moved towards the patient from the end of the rule and the patient asked to state when the print becomes blurred (push-up method). The monocular near point of accommodation is also assessed to determine any difference between the monocular and binocular accommodative states. The results are recorded in centimeters and repeated three times both binocularly and unioocularly to assess the effect of fatigue.

The examiner is able to identify if the level of accommodation is adequate for the patient's age from the age scale and the amount of accommodation exerted at a given distance can be determined from the dioptre scale. The patient's blur point is an indication of the amplitude of accommodation.

Amplitude of accommodation can also be assessed using trial lenses. The patient is seated 6 m from a Snellen test type wearing the appropriate prescription and with one eye occluded. Whilst viewing the smallest print that can be seen clearly, minus lenses are introduced in steps of 0.5–1 dioptre until the print blurs. The sum of the strength of lens through which the patient can no longer see clearly, is equivalent to the amplitude of accommodation. Amplitude is correlated to age.

The *accommodative facility* is a measure of how fast a patient can report that clarity has been restored following a rapid change of focus. Assessment involves stimulation followed by relaxation of accommodation using –2.00 and +2.00 dioptres sphere (DS) lenses (Fig. 4.24). One cycle equals accommodative stimulation and relaxation and the test is measured in cycles per minute. The assessment is



Figure 4.24 Flipper lenses.

performed binocularly and unioocularly. Normal facility for adults is 8 cycles/minute binocularly and 11 cycles/minute unioocularly (Henshall & Rowe 2002). For children, this is 5 cycles/minute binocularly and 7 cycles/minute unioocularly.

Convergence

Convergence is a disjunctive movement in which both eyes rotate inwards. There are four types of convergence:

1. *Tonic*, which is due to the tone of the extraocular muscles initiating movement from the anatomical position of rest.
2. *Accommodative*, which is initiated by the stimulus of accommodation.
3. *Fusional*, which is initiated by a fusional stimulus. This is an involuntary vergence movement to maintain binocular single vision.
4. *Proximal*, which is induced by the awareness of a near object.

All four components of convergence contribute to the total amount of convergence. One-third of convergence relates to tonic, fusional and proximal convergence. Two-thirds is accommodative.

Relative convergence is the degree of convergence produced relative to the amount of accommodation exerted and may be positive (increased) or negative (decreased) in order to maintain binocular single vision. The 'metre angle' is one

unit measurement for the amount of convergence present. One metre angle equals the amount of convergence required for each eye to fixate an object, located at a distance of 1 m, in the median plane having moved from fixating a distant object.

The amount of convergence is calculated by using the formula: $1/d \times \text{IPD}$ where d is the distance and IPD is interpupillary distance. Thus, a 6-cm IPD requires 6 dioptres of convergence for a fixation distance of 1 m.

Measurement of convergence

The *near point of convergence* is assessed objectively using either a fixation target or the fixation target on the RAF rule. Normal convergence is to 6 cm. The fixation target or the RAF rule is held in the primary position. The target is gradually moved, from at least 1/3 m away from the patient, slowly and steadily towards the patient's eyes. The patient is instructed to maintain fixation on the target and states when the target blurs, jumps or becomes double. The examiner notes the point at which this occurs by observing the break of convergence in the patient's eyes. Pupil dilatation may be observed at this stage, which is an additional indicator of convergence failure.

Convergence gives an indication of the strength of binocular single vision (Capobianca 1952).

AC/A ratio

This is the amount of accommodative convergence that is induced by each dioptre of accommodation exerted and is measured in prism dioptres. It is a fixed relationship, so that an equal amount of convergence occurs for each dioptre of accommodation. This is inborn and remains virtually unchanged from childhood to at least presbyopic age, provided that the stimulus to accommodate is kept within narrow limits (Breinin & Chin 1973). The normal range is 3–5:1 (Ogle *et al.* 1966). The amount of accommodative convergence exerted depends on the interpupillary distance and distance of the object from the eyes.

The AC/A ratio can be modified permanently by surgery of the extraocular muscles and temporarily by drugs and lenses that reduce the accommodation requirement and hence the resulting accommodative convergence.

Methods of measurement of AC/A ratio

Gradient method

1. Near measurement, using convex lenses. A prism cover test is performed for near fixation with and without convex lenses (usually +3.0 DS).
2. Distance measurement, using concave lenses. A prism cover test is performed for distance with and without concave lenses (usually -3.0 DS).

Calculation:

$$\text{AC/A ratio} = \frac{(\text{PCT with accommodation exerted}) - (\text{PCT without accommodation exerted})}{\text{Amount of accommodation exerted}}$$

Heterophoric method A prism cover test is performed for near and distance fixation and the interpupillary distance is measured. A positive sign is used for eso-deviations and a negative sign is used for exo-deviations.

Calculation:

$$\text{AC/A ratio} = \text{IPD(in cm)} + \frac{(\text{Near PCT}) - (\text{Distance PCT})}{\text{Amount of accommodation exerted}}$$

Graphic method The angle of deviation is measured for distance fixation on the synoptophore or using the prism cover test. This is repeated using concave lenses, starting with -1.0 DS and increasing in 1-dioptre steps to -4.0 DS if possible. A Snellen test type chart is used and the patient is instructed to fixate the same line of letters at each measurement.

Calculation: A graph is plotted showing change in convergence in prism dioptres against the change in accommodation in dioptres.

The stimulus AC/A ratio is the degree of convergence elicited in relation to the stimulus of accommodation and is measured clinically by gradient, heterophoric and graphic methods. The response AC/A ratio is the degree of convergence elicited in response to the physical accommodation exerted.

The AC/A ratio measurement assumes that the accommodation changes when looking through the minus or plus lens is equivalent to the power of the lens. However, a lag of accommodation may occur where the level of accommodation is less than the stimulus power (with minus lenses) or there is incomplete relaxation of accommodation (with plus lenses). This leads to higher AC/A ratio results. The response AC/A ratio has been found to be higher than the stimulus ratio, which is explained by accommodative lag with minus lenses (Gratton & Firth 2010).

When measuring the AC/A ratio, it is important to control accommodation by choosing the smallest target size. Presence of accommodation can be checked by observing pupils for miosis. Fusional convergence should be eliminated by complete dissociation.

Uses of AC/A ratio

- Diagnosis of convergence excess type intermittent esotropia (see Chapter 6).
- Differential diagnosis of true and simulated intermittent distance exotropia (see Chapter 6).

CA/A ratio

This is the extent of accommodation resulting directly from convergence. The CA/A ratio assesses whether a known amount of convergence results in excessive or defective convergence accommodation. There is a reciprocal relationship between the CA/A and AC/A ratios in that where one ratio is high, the other is low (Schor 1988).

The CA/A ratio is calculated by comparing the refractive error of each eye before and after convergence is induced by a base-out prism. The ratio is expressed as the amount of convergence accommodation per metre angle of convergence with a normal expected value of 0.5–1 m angle.

Retinal correspondence

Tests that are used to investigate retinal correspondence are often dissociative and usually subjective and include the following:

- *Diplopia based:* For example, Bagolini glasses and Worth's four light test. These tests rely on the subjective localisation of a single object whose image is formed on the retinal elements stimulated.
- *Haploscope-synoptophore:* Two dissimilar objects are presented in the binocular field of view and the patient is required to superimpose the two images.

Bagolini glasses test

This is a minimally dissociative diplopia based test that investigates the presence of retinal correspondence. The test may be used in Lorgnette form or separately in a trial frame. Each glass is made of plain glass incorporating fine parallel striations placed before both eyes with the striations at 45° and 135°. A spotlight viewed through the glass produces a line image seen perpendicular to the striations.

The patient is asked how many line images and spotlights are seen and what form the lines make (Fig. 4.25). Possible results include the following:

- One spotlight with two line images forming a cross through the middle of the light indicates normal or abnormal retinal correspondence (Fig. 4.26a). A gap in one of the lines around the light indicates a suppression scotoma (Fig. 4.26b).
- Two lights with the two lines displaced indicate a manifest deviation with diplopia. Normal retinal correspondence is present if a cross response is achieved when the angle of deviation is corrected (Fig. 4.26c).
- One light with one line image indicates suppression.



Figure 4.25 Bagolini glasses.

Worth's four light test

This test is used to investigate the presence of retinal correspondence. The subject may be tested at 1/3 m and 6 m. It consists of four circular lights on a black/grey background – one red, one white, two green, which are set in a diamond pattern and illuminated from within. The patient views these lights using red and green glasses, with the red filter before the right eye.

This is a dissociative diplopia based test and uses complementary colours. The white light is common to both eyes and therefore provides a fusional stimulus. The patient views the lights and is asked to state how many are seen (Fig. 4.27).

Possible results include the following:

- *Four lights (one red, two green and one light that is a mixture of red and green):* It indicates normal or abnormal retinal correspondence.
- *Five lights:* It indicates a manifest deviation with diplopia. The lights are displaced according to the type of deviation present. Two red lights on the left and three green lights on the right sides are seen with exo-deviations and with eso-deviations; the green lights will be on the left and the red on the right side. Normal retinal correspondence is present if four lights are seen when the deviation is corrected.
- *Three or two lights:* It indicates suppression. Three green lights indicate right suppression and two red lights indicate left suppression.

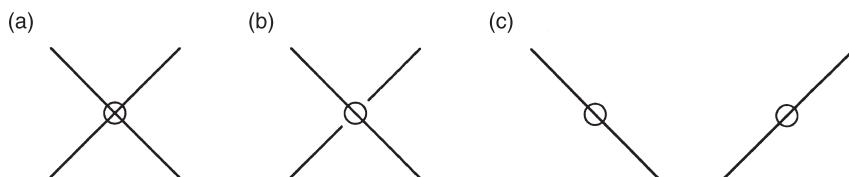


Figure 4.26 Results with Bagolini glasses.

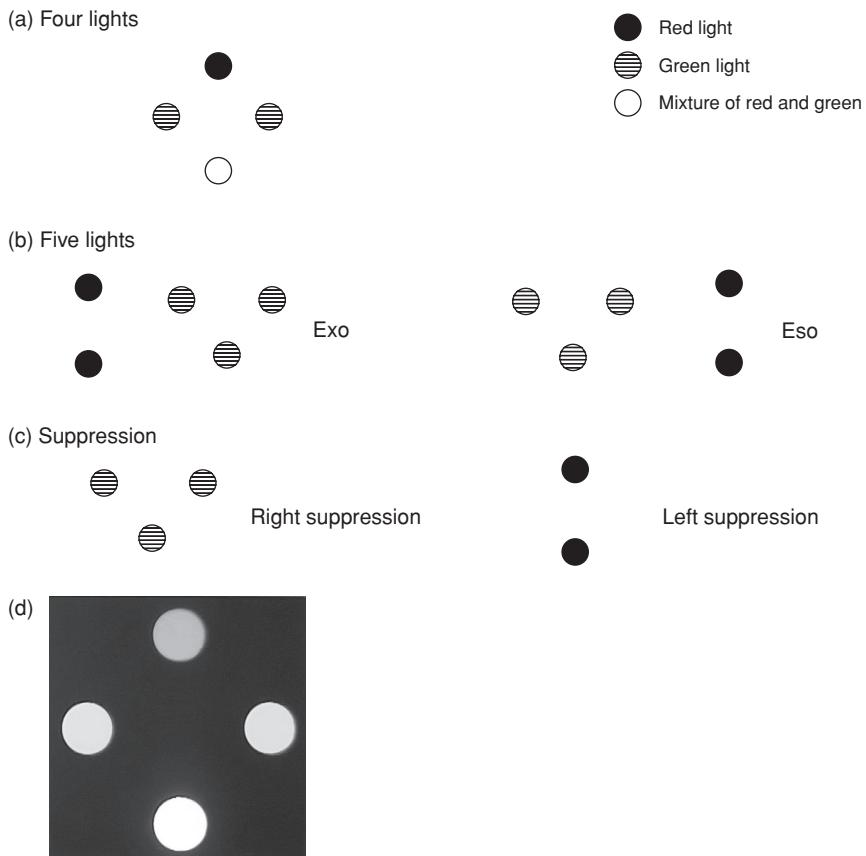


Figure 4.27 Worth's four lights test.

After-image test

When the eye is exposed to a bright flash of light, the subjective sensations last longer than the stimulus itself. This sensation is the after-image. The after-image test uses fovea/foveal projection to demonstrate the relationship between the foveal projection of both eyes. There is bifoveal stimulation but not simultaneous stimulation. It is a test of retinal correspondence because it compares the retinal areas that have the same visual direction during binocular single vision (Bielchowsky 1937).

An after-image is presented to each eye in turn in the form of a linear bright light, either horizontal or vertical, with a central fixation target on each linear light. The horizontal line image is presented to the fixing eye first, followed by a vertical image to the deviating eye (this minimises suppression).

An after-image may be produced by an electronic flash gun or by using the synoptophore. The flash stimulus produced by an electronic flash gun is presented to each eye in turn and the subjective responses are recorded.

The synoptophore may be used to produce an after-image when an autofocus device has been incorporated into the instrument. After-image slides of the line images are required. Controls are set at zero and the opal filters are removed. The slides are inserted with the horizontal image before the fixating eye and the vertical image before the deviating eye. The autofocus is adjusted to rapid and simultaneous settings and the light intensity is increased to maximum strength.

The tube is switched on before the fixing eye for 10 seconds and then switched off. This is followed by the same procedure before the deviating eye. The opal filters are then replaced, the slides removed and the autofocus is switched on. The patient looks into the synoptophore or at a blank wall, and describes the subjective after-image.

Possible results include the following:

- Foveae that project to the same point in space indicate normal retinal correspondence.
- A displaced after-image is produced by the deviating eye in cases of manifest strabismus because the foveae project to different points in space. This includes cases with abnormal retinal correspondence as the stimulus is to the fovea and not the pseudofovea in the deviating eye.

Prism adaptation test

This test assesses the state of binocular function in addition to obtaining the maximum angle of deviation (Kassem 2008). The angle of deviation is overcorrected by 5–10 prism dioptres with fresnel prisms. The patient's motor response is observed after a period of at least 24 hours. Possible results include the following:

- The visual axes may assume a straight position or a microtropia, demonstrating binocular single vision.
- The visual axes may resume their original position, which indicates abnormal retinal correspondence.
- The visual axes remain in the overcorrected position indicating absence of any binocular function. Where the angle increases, the prism strength increases further until the deviation stabilises. This stable angle represents the true angle of deviation. A 'non-responder' demonstrates no binocular potential, an exo-shift and exo-deviations greater than 60 prism dioptres.

Vertical prism test

In cases of manifest horizontal strabismus with suppression, a 10 or 15 prism dioptre vertical prism may be used to displace an image onto the superior or inferior retina and produce diplopia. A second horizontal prism is used to place the images in vertical alignment.

Normal retinal correspondence is present if the strength of the second prism is equivalent to the objective measurement of the angle of deviation. If the second prism strength is equivalent to the patient's subjective angle of deviation, then abnormal retinal correspondence is present. If the patient is unable to appreciate the images in vertical alignment, then suppression is present and retinal correspondence cannot be determined.

Synoptophore

Simultaneous perception (SP) slides are used. The method is described further later in the chapter.

Botulinum toxin A

Botulinum toxin A can be used to decrease the angle of deviation in order to assess the potential for post-operative binocular single vision. In esotropia, the medial rectus is injected and in exotropia the lateral rectus is injected with botulinum toxin to paralyse the muscle's function. The eye takes up a straighter or overcorrected angle post injection. When the eye is in a 'straight' position, the state of retinal correspondence and potential for binocular single vision can be assessed.

Fusion

Sensory fusion is the ability to perceive simultaneously two images – one formed on each retina – and interpret them as one. It has been advocated that sensory fusion may be assessed using Bagolini glasses and Worth's lights. However, these tests are only an assessment of retinal correspondence indicating the ability or inability of the patient to superimpose images. Sensory fusion may be assessed on the synoptophore or the Sbrisac bar can be used to test sensory fusion (Hocking & Gage 2002). The mean strength at near fixation using the Sbrisac bar was 11.01 ± 3.37 and for distance fixation was 11.25 ± 3.06 . Thus, sensory fusion measured with the Sbrisac bar can be tested at one distance only.

Sensory fusion assessed using the Sbrisac bar with Bagolini glasses incorporated to aid patient interpretation and understanding has been recommended as a clinical assessment (Knowles & Griffiths 2003). The mean filter strength at near fixation was 13.91 ± 2.13 when fixing with the non-dominant eye and 14.46 ± 1.11 when fixing with the dominant eye. Therefore, it is important to specify which eye is tested as dominance has an effect on outcome.

Motor fusion is the ability to maintain sensory fusion through a range of vergence movements. The strength of binocular single vision is indicated by measuring the range of motor fusion. Vergence adaptation is important for achievement of binocular vision. The motor fusion range is a response to disparity by base-out and base-in prisms (convergence and divergence) and represents fast fusional vergence (step fusion). The ability to sustain these vergences is maintained by the slow vergence system (tonic fusion).

Motor fusion may be measured:

1. qualitatively by:
 - cover test recovery;
2. quantitatively by:
 - prism fusion range;
 - prism reflex test;
 - binocular visual acuity;
 - controlled binocular acuity;
 - bar reading;
 - synoptophore.

Cover test recovery

The recovery movement is assessed on removal of the cover from either eye as this movement denotes the presence of binocular single vision and therefore fusion. Full recovery whereby the eyes move back to a straight position indicates normal binocular single vision.

Prism fusion range

If a base-out prism is placed before one eye, the target image is displaced in the direction of the prism apex. In order to maintain binocular single vision, the eye turns inwards by a corrective amount. The strongest base-out prism through which binocular single vision can be maintained marks the limit of the patient's convergent fusion range.

When assessing the divergent fusion range with base-in prisms, the target image is displaced towards the prism apex and the eye turns outwards by a corrective amount. The strongest base-in prism through which binocular single vision can be maintained marks the limit of the patient's divergent fusion range (Figs. 4.28 and 4.29).

Phasic fusion is measured using stepped changes with prism bars. Tonic fusion is measured using a smooth transition with Risley prisms. Differences in fusional amplitude between tonic and phasic methods may be found and may explain symptomatic binocular vision for some patients. It is important to ensure the prism bar is held in the correct position (Prentice position, position of minimal deviation or frontal plane position) as incorrect use may overestimate the measurement of prism fusion range (Bath & Firth 2007).

Horizontal fusion range

Latent deviation

The patient fixates a target at 1/3 m or 6 m held in the primary position.

A horizontal prism bar is placed before one eye and starting with the weakest prism strength, is moved through the range until the patient is no longer able to



Figure 4.28 Risley prism (a): Prism bars and loose prisms (b).

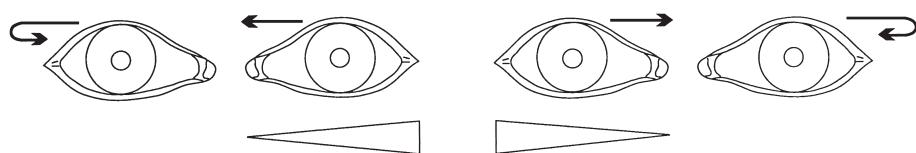


Figure 4.29 Response to overcome a base-out prism. A base-out prism is placed before the left eye. Both eyes make a conjugate movement to the right and the right eye makes a refixation movement to the left. When the prism is placed before the right eye, a similar response is obtained, with a conjugate movement of both eyes to the left followed by a refixation disjunctive movement of the left eye to the right.

maintain binocular single vision. This can be determined by observing the patient's eyes to ensure that the prism is overcome. The strength of prism at which the patient no longer makes a fusional movement to overcome it marks the limit of the fusional range. The eyes are seen to diverge and the patient may appreciate diplopia. If unsure of the break point, Bagolini glasses may be used as a control. The cross response will split into two separate lines on break of fusion. When fusion breaks, allow time to refuse the images if the patient is able. Once fusion is lost, decrease the prisms until the patient is able to regain fusion; this is usually at a level 4–6 prism dioptres lower than the break point. If the level is more than this, there may be an underlying problem with compensation. The blur point measures the limits within which accommodation can clear the image of the fixation target in spite of increased convergence. The amount of fusional convergence that can be elicited between the blur point and break point represents the absolute convergence.

The test is performed with the prisms held base out and base in. On increasing the base-out prisms (convergence range), the patient may appreciate a decrease in size of the image (micropsia). On increasing the base-in prisms (divergence range), the patient may appreciate an increase in the size of the image (macropsia). Quite often, the patient will be aware of blurring of the image before the image actually splits on break of fusion. This is termed the blur point of fusion (Narbheram & Firth 1997) and is an important indicator of whether accommodative vergence is being used to aid fusional vergence.

The normal fusional range is 35/40 prism dioptres base out and 16 prism dioptres base in on near testing, and 16 prism dioptres base out to 8 prism dioptres base in on distance testing (Berens 1927, Scobee & Green 1948, Tait 1949, Crone *et al.* 1975, Kertesz 1978, Guyton 1988, Sharma 1992).

Normal adults can fuse 8° of cyclodisparity using sensory fusion (Guyton 1988).

Choice of target size is important with smaller fusion ranges documented for central (0.0 logMAR) targets compared to peripheral (1.0 logMAR) target sizes. A normal fusion range at near fixation is base-out 25 to base-in 10 prism dioptres and base-out 35 to base-in 12 prism dioptres for central and peripheral targets respectively. At distance fixation, the normal range is base-out 16 to base-in 6 and base-out 25 to base-in 6 prism dioptres for central and peripheral targets respectively (Rowe 2010). Alterations in fusional vergence may also be seen if prisms are held before the non-dominant or dominant eye (Hainey *et al.* 1999) with a slightly increased base-out range when the prism is placed before the non-dominant eye.

Manifest deviation

The angle of deviation is first corrected with prisms that are then gradually altered (increased and decreased) in strength to assess the motor fusion range. Bagolini glasses may be used as a control to ensure that the patient does not suppress one image.

Vertical fusion range

This is an important test for the differential diagnosis of long-standing decompen-sating fourth nerve palsy and acquired fourth nerve palsy. Base-down prisms

are used to assess supravergence and base-up prisms for infravergence. The normal range is 3 prism dioptres base down and 3 prism dioptres base up (Sharma & Abdul-Rahim 1992). Vertical fusion is slightly greater at near than distance fixation but not significantly different (Ulyat *et al.* 2004).

Prism reflex test

Babies and young children who are unable to cooperate with a prism fusion range test may be assessed using single base-out prisms. Using a 20 dioptre prism base out, the prism is placed before either eye in turn with the examiner looking for a movement of the eyes to overcome the effect of the prism and looking at the rate of recovery and extent of recovery. If the patient does not overcome the 20 dioptre prism, the test should be repeated with lower strength prisms down to 10 dioptre prism before recording absence of fusional movements.

Fusion vergence facility

This is the ability to converge and diverge the eyes rapidly in response to base-out and base-in prisms. It is the speed with which a patient can recover fusion or sustain binocular vision in the presence of rapid changes in vergence demand. A reduced fusion facility demonstrates fatigue of binocular vision. This can occur despite a normal motor fusion range but may be symptomatic because of a fatigue effect.

Fusion facility is assessed using flipper prisms containing a base-out and base-in prism held at 40 cm, for example base-out 40 and base-in 12 prism dioptres or base-out 12 and base-in 8 dioptres. This induces jump vergence to both prisms. One cycle represents a response to overcome to the effect of both the base-out and base-in prisms consecutively. The test is recorded in cycles per minute.

Binocular visual acuity

This test is used to assess the strength of binocular single vision. The test is important as patients can exert accommodation to control their deviation. Therefore, during testing, intermittent occlusion acts as dissociation and control can break at a level of visual acuity less than each monocular visual acuity.

The patient is required to read decreasing sizes of print whilst maintaining binocular single vision at both 1/3 m and 6 m. The examiner must record the level of monocular visual acuity first. The patient is then instructed to read slowly down the letter chart. The examiner observes the eyes for a manifest strabismus. A cover/uncover test is performed for each size of letter to determine if binocular single vision is still present.

If a latent deviation becomes manifest, the binocular visual acuity is recorded for the previous line of letters. The maximum level of binocular visual acuity is equivalent to the weakest monocular visual acuity.

Controlled binocular acuity

The strength of binocular single vision is assessed by this method in the same manner as for binocular visual acuity. However, intermittent occlusion as a dissociation is not used during the test (Firth & Davis 2009).

The patient is required to read decreasing sizes of print whilst maintaining binocular single vision at both 1/3 m and 6 m. The examiner must record the level of monocular visual acuity first. The patient is then instructed to read slowly down the letter chart. The examiner observes the eyes for a manifest strabismus.

If a latent deviation becomes manifest, the binocular visual acuity is recorded for the previous line of letters. The maximum level of binocular visual acuity is equivalent to the weakest monocular visual acuity.

Synoptophore

This can be used to assess the presence of sensory fusion. Motor fusion is assessed by converging and diverging the tubes. This will be further discussed later in the chapter.

Diagnostic occlusion

Occlusion of one eye for at least half an hour is recommended to determine if strong fusional vergence is responsible for control of a deviation. Monocular diagnostic occlusion will relieve the fusional control and directly following removal of the occlusion, the deviation may be seen to have increased considerably in angle. This angle can be measured by the prism cover test and it is essential that following removal of the occlusion the patient does not have binocular viewing prior to completing the angle measurement. The final angle measurement can be regarded as a truer estimate of angle of deviation.

Stereopsis

Depth may be perceived in two forms: binocular disparity (stereopsis) and monocular depth clues in the absence of binocular single vision.

Stereopsis is the perception of the relative depth of objects on the basis of binocular disparity, which reflects the slight difference in images presented to each eye.

Stereoacuity is the angular measurement of the minimal resolvable binocular disparity, which is necessary for the appreciation of stereopsis. The ability to demonstrate stereopsis indicates the presence of retinal correspondence, the normal level of stereopsis being 40'' with normal equal visual acuity in each eye. The level of stereoacuity decreases with reduction in visual acuity (Oguz & Oguz 2000) and there is an age-related deterioration in stereoacuity with a linear correlation between age and stereopsis threshold (Zaroff *et al.* 2003).

Principle of random dot stereograms

Random dot stereograms comprise images, formed of dots that are displaced in relation to each other. The space created by this is filled randomly by dots, those seen by one eye having no relation to those seen by the other eye. This produces disparity of viewing as both eyes will see different dots. With stereopsis, these disparate images are fused and depth is appreciated.

Random dot tests lack monocular clues but are more difficult (Simons 1981). The inability to perform on random dot may be due to a crowding effect from closely placed random dots.

Random dot stereogram tests include the following:

- Lang stereotest
- TNO stereotest
- Synoptophore

Lang stereotest

This is based on the use of random dot stereograms and cylindrical grating placement (panography) spaced at 24 cylinders per centimeter. Beneath each cylinder, there are two strips of an image, one seen by the right eye and the other seen by the left eye. The two images are fused and the disparity produces stereopsis (Lang 1983). The first Lang stereotest consists of three images – cat, car and a star – and the second Lang stereotest consists of four images – the star, which is a control image and can be seen in the absence of stereopsis, the car, the elephant and the moon, which are seen in the presence of stereopsis up to 200" (Fig. 4.30).

TNO stereotest

This is based on the use of random dot stereograms. Computer-generated random dots are printed as red and green analyphs. Red and green glasses are worn, which are complementary colours to the red and green random dots used. It consists of three gross stereoplates, a suppression plate and three graded plates with segmented circle shapes, which give a result of 480"-15" (Okuda *et al.* 1977). This is considered the most accurate test for stereopsis as there are no monocular clues (Fig. 4.31).

Synoptophore

There are two different groups of slides in use:

1. Graded stereoscopic slides, which contain two dissimilar images. Each has a control but there is an inbuilt disparity between the images that when fused will produce gross stereoscopic vision.
2. Braddick slides consist of pairs of random dots, each slide stimulating slightly disparate retinal points resulting in depth disparity.

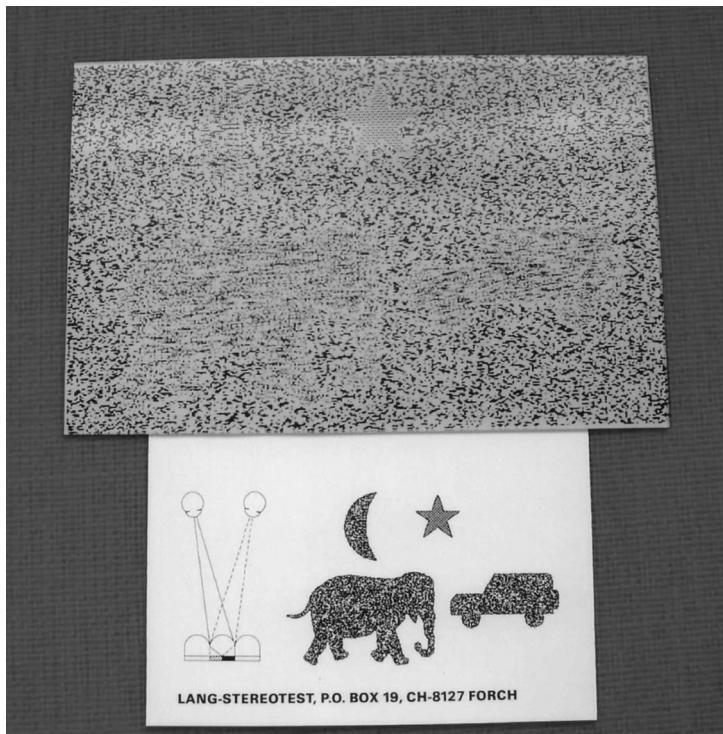


Figure 4.30 Lang stereotest.

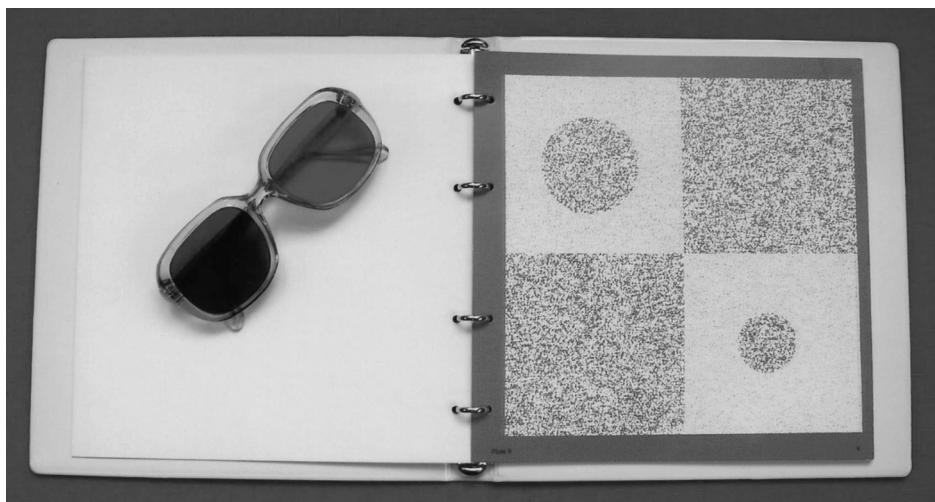


Figure 4.31 TNO stereotest.

Principle of linear polarisation

Polaroid filter glasses are worn and a vertical image of a light source is seen through one filter and a horizontal image of the light is seen through the other. The targets are presented as vectographs and consist of two polarised surfaces with a slight crossed disparity between them. The disparity produced by either eye results in stereopsis.

Linear polarisation stereotests include the following:

- Titmus/Wirt stereotest
- Randot stereotest

Titmus/Wirt stereotest

This is based on linear polarisation and consists of two plates. One plate contains the image of a fly and three rows of animals. Gross stereopsis is demonstrated if the wings of the fly are perceived as standing out from the body of the fly. The image of one animal in each row also appears to stand out and stereopsis of up to 100" may be achieved. The second plate consists of nine boxes each containing four circles. One circle is displaced to produce disparity, and stereopsis of up to 40" may be achieved (Fig. 4.32).

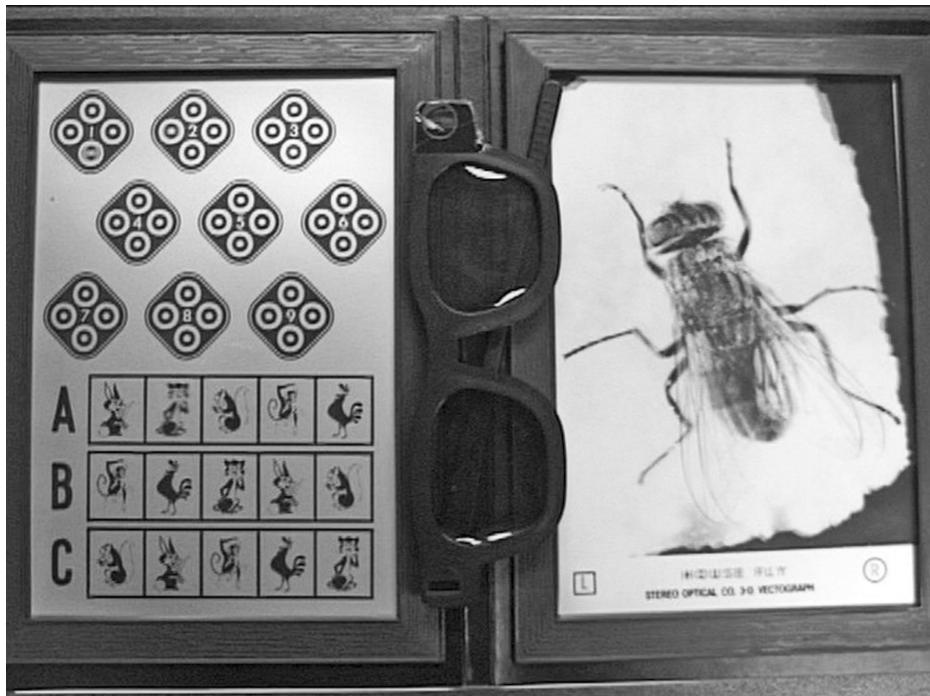


Figure 4.32 Titmus/Wirt stereotest.

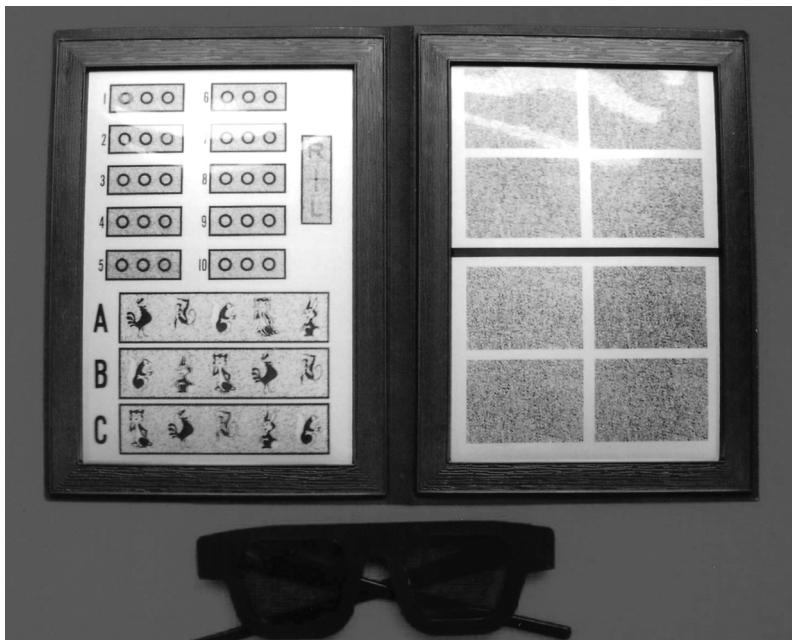


Figure 4.33 Randot stereotest.

The Titmus stereotest has limitations due to monocular clues. False positive responses with Titmus are reported (Simons 1974). The most obvious of these is lateral displacement of targets (Cooper & Warshowsky 1977). True stereoacuity may be confirmed by rotating the test target 180°. The previously elevated target should sink below the plane of the book.

Randot stereotest

The near test is based on linear polarisation and consists of two plates containing random forms. The perception of random dot shapes gives a result of 500''–250''. Appreciation of depth with the animals and circles gives a result of 400''–20'' (Fig. 4.33). Completion rate is excellent with this test with normative values varying according to age: 100'' for age 3, 70'' for age 4, 50'' for age 5, 40'' for age 6 and 45'' for age 7 (Kulp & Mitchell 2005).

A distance Randot stereotest is available for assessment of stereoacuity at 6 m viewing (Wong *et al.* 2010).

Lang two pencil test

This is a test for gross stereopsis. The patient attempts to place a pencil on top of one held by the examiner. Patients who possess stereopsis will find it easier to perform the test with both eyes open than monocularly (Fig. 4.34). The two-pencil test gives stereopsis estimates of 3000''–5000'' (LaRoche 1982).

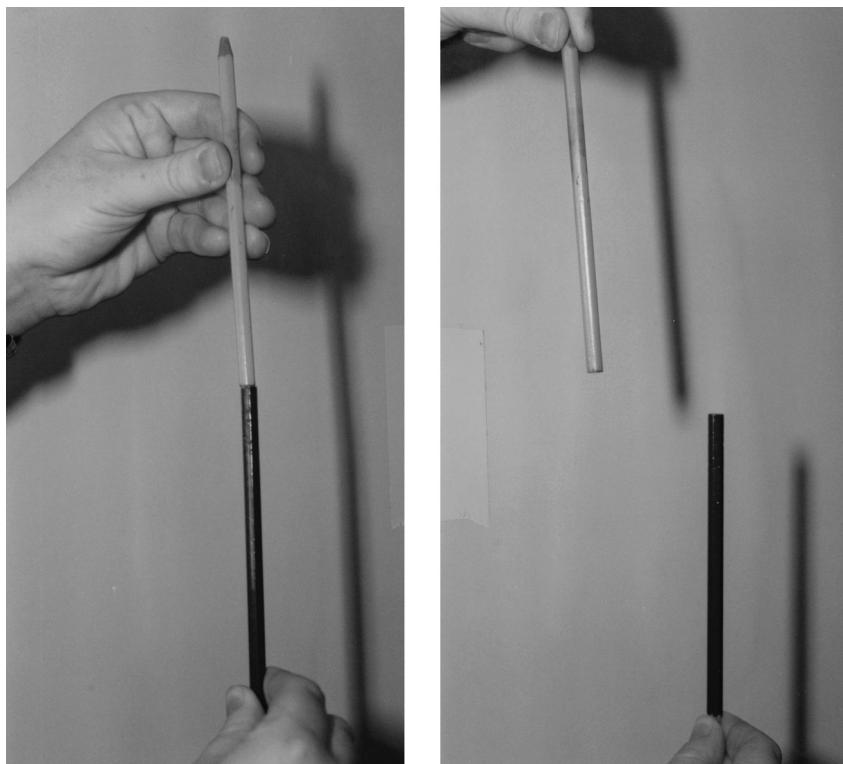


Figure 4.34 Lang two pencil test.

Frisby stereotest

The near Frisby stereotest uses random shapes with displacement and consists of three plastic plates of different thickness: 6 mm, 3 mm and 1 mm. On each plate, there are four squares produced by random shapes printed on one side and on the other, corresponding to one of the squares, a disparity is created by printing a circle with random shapes. This gives a real-time 3 dioptres appearance. The disparity is affected by the thickness of the plate and the distance at which the plate is held from the patient. When held at 40 cm, stereopsis of 340", 170" and 55" can be achieved with the 6, 3 and 1 mm plates, respectively. The Frisby stereotest is very useful for assessing stereopsis in very young children and patients with easily dissociative strabismus (Fig. 4.35).

The FD2 (Frisby-Davis) stereotest is a measure of distance stereopsis and consists of four translucent shapes, one mounted in each quadrant of a back-illuminated display box (Fig. 4.36). The test uses real depth and the shapes are moved in relation to each other. The patient identifies which shape appears closer to them and a stereopsis range of 5"-50" can be achieved (Davis *et al.* 2002).



Figure 4.35 Frisby stereotest.

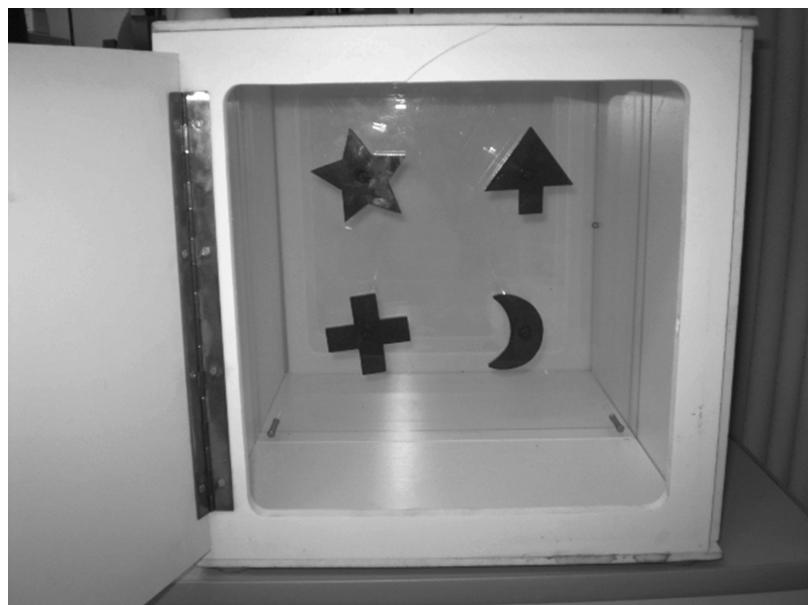


Figure 4.36 FD2 stereotest.

Suppression

The presence of suppression can be detected by using Bagolini glasses, Worth's lights, the Synoptophore and the 4 dioptre prism test. Various stereopsis tests also incorporate a suppression test plate, for example TNO. The area and density of suppression may also be measured.

Area of suppression

The area of suppression may be plotted using the synoptophore with SP slides. The tube is locked at zero before the fixating eye. The tube is rotated horizontally and then vertically before the suppressing eye and the angles at which the image of the deviating eye disappear and reappear are recorded. This forms an area of suppression.

Different sizes of graded SP slides may also be used to assess a suppression scotoma. The patient will only be aware of both slides if the picture in front of the suppressing eye is large enough to lie outside the suppression scotoma. It follows that the larger the slide required in front of the deviating eye to enable it to be seen, the larger the size of the suppression scotoma.

The area of suppression may also be assessed with prisms in the same way that post-operative diplopia is assessed. Prisms are placed before the suppressing eye and gradually increased in strength until diplopia is noticed. Prisms are introduced base out, base in, base up and base down to assess the complete area of suppression.

Depth/density of suppression

A Sbisa (Bagolini filter) bar (Fig. 4.37) is used to investigate the depth of suppression in patients with amblyopia approaching the age of 7 years who require treatment (Price & Auld 2008). If suppression is very dense, the decision may be taken to occlude carefully. The Sbisa bar consists of a series of red filters of increasing density from very pale to very dark. This decreases the illumination in front of the fixating



Figure 4.37 Sbisa bar.

eye to induce fixation with the deviating eye and therefore diplopia. The patient fixates a spotlight held at 1/3 m in the primary position. The Sbisa bar is introduced before the fixating eye commencing with the palest filter. The light will then appear pale red. The filter strength is increased until the patient sees one white and one red light as a result of diplopia or one white light as a result of swapped fixation or the light stays red with continued consistent fixation. The strength of filter at which this occurs represents the density of suppression.

Post-operative diplopia test

This test is used to investigate the likelihood of a patient experiencing diplopia post-operatively. It may be performed on any patient over the age of 7 years who is being considered for cosmetic strabismus surgery. It investigates the area of suppression and therefore the possible occurrence of post-operative diplopia if surgery were to result in ocular alignment in which images fall outside the suppression area. At this stage, the patient would be unable to resuppress and would appreciate intractable diplopia.

The patient fixates a spotlight at 1/3 m and prisms are introduced initially at the lowest strength and then gradually increased in order to first undercorrect, then correct the angle of deviation and eventually overcorrect it. For each strength of prism, the patient is asked to state what is seen, that is, single image or diplopia, and the test is then repeated at 6 m. Red and green glasses may be used to facilitate the appreciation of diplopia, but in clinical practice the test is often performed without these glasses as this is more realistic of the patient's everyday viewing conditions (Gray *et al.* 1996). Possible results include the following:

- Should the patient continue to see one light, it is unlikely that intractable diplopia would pose a problem post-operatively.
- Diplopia may be appreciated when the deviation is overcorrected and this would alert the surgeon not to overcorrect the deviation.
- Diplopia may be appreciated intermittently when undercorrected and this may be a contraindication to surgery.

Should the patient appreciate diplopia, it is important to assess whether this may be ignored, particularly if vision in one eye is poor. When diplopia is appreciated, the patient is asked whether they are capable of ignoring or resuppressing the second image and how easy this is to achieve. Where it is relatively easy to ignore the second image, the patient may feel that the cosmetic appearance warrants the risk of diplopia post-operatively.

Where further information regarding post-operative diplopia is required, use of botulinum toxin may be considered to align the eyes temporarily. A smaller percentage of patients report post-operative diplopia following use of botulinum

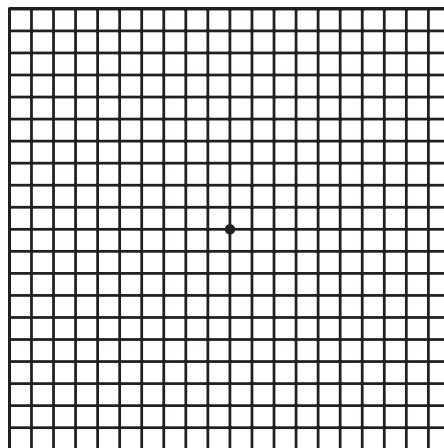


Figure 4.38 Amsler chart.

toxin versus use of prisms and patients are able to determine in ‘real life’ situations whether they are able to cope with diplopia or not (Khan *et al.* 2008).

Amsler charts

These may be used to plot central and paracentral scotomas. The chart usually consists of a square grid of black lines on a white background. The patient describes or draws how the grid appears, thus demonstrating the scotoma (Fig. 4.38).

Synoptophore

This is a haploscopic device in which each eye looks through a separate tube, viewing a picture on a slide on a plane mirror. The strength of the lenses within the eye pieces are +6.5 DS, and as a result it is considered to be a distance test (Figs. 4.39 and 4.40).

Slides

There is a range of slides. The sizes of the pictures on the slides subtend visual angles of differing degrees at the nodal point. Foveal SP is the ability to perceive simultaneously and to superimpose two dissimilar images subtending angles not greater than 1° at the nodal point. Macular SP is the ability to perceive simultaneously and to superimpose two dissimilar images subtending angles greater than 1° but not greater than 3° at the nodal point. Paramacular SP is the ability to perceive simultaneously and to superimpose two dissimilar images subtending angles greater than 3° but not greater than 5° at the nodal point. Peripheral SP is the ability

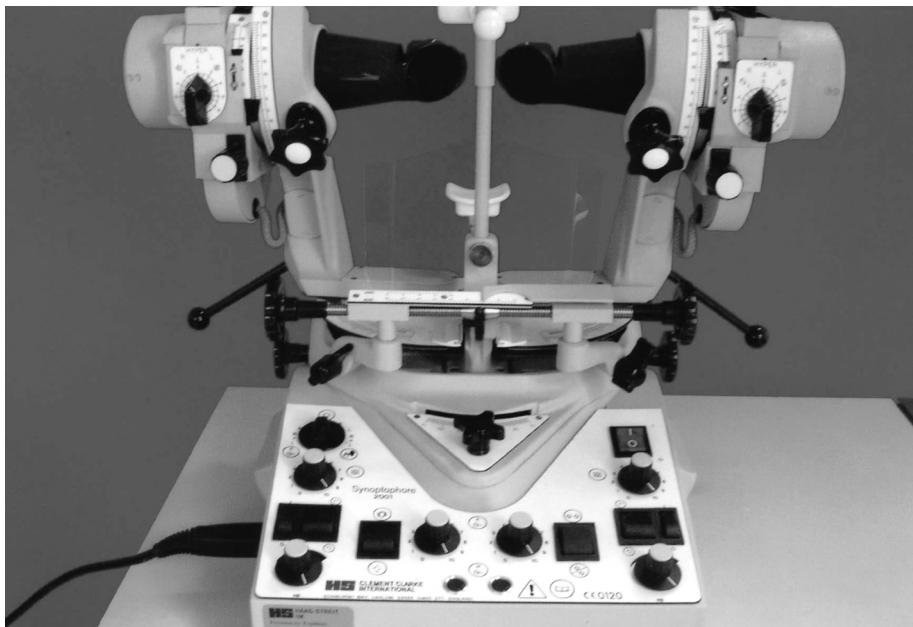


Figure 4.39 Synoptophore.

to perceive simultaneously and to superimpose two dissimilar images subtending angles greater than 5° at the nodal point:

- SP slides are red in colour and are used to measure IPD, angle of deviation and assessing SP.
- Fusion slides are green in colour and are used to measure sensory and motor fusion.
- Stereopsis slides are yellow in colour. Sizes include gross, detailed or Braddick.

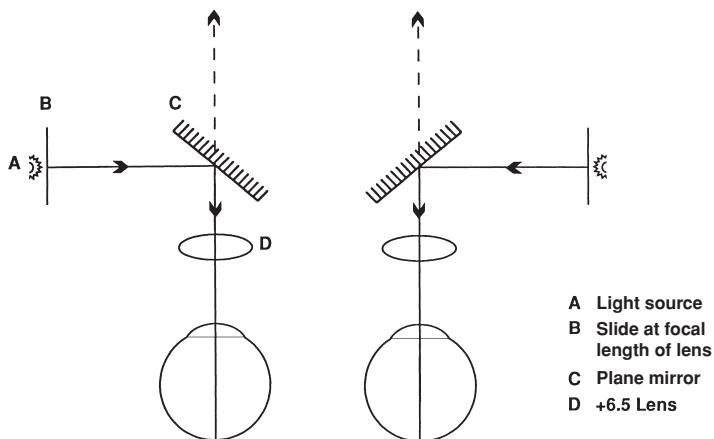


Figure 4.40 Optics of the synoptophore.

Uses of the synoptophore

Measurement of interpupillary distance

The *interpupillary distance* (IPD) is measured. The interpupillary distance is the distance between the two pupil centres. This procedure is performed on all patients so that they are aligned properly such that each eye is looking centrally down each tube. The average for an adult is 65 mm when the eyes are fixing in the distance, the average for a child is 50 mm. The set-up and measurement of IPD must be correct before any other measurements may be made.

Measurement of the angle of deviation

The test may be performed either objectively or subjectively, both with and without spectacles, fixing either eye, in nine cardinal positions. The tubes are rotated horizontally and/or vertically by up to 25° (elevation, adduction and abduction) and 35° (depression) for this purpose and horizontal, vertical and torsional angles of deviation are obtained.

SP slides appropriate to the visual acuity are used. However, Maddox slides are used for measurement of incomitant deviations (Fig. 4.41). The tube is locked before the fixing eye and an alternate cover test performed by extinguishing the light in each tube to act as an occluder. With eso-deviations, the tubes are converged before the deviating eye, and with exo-deviations, the tubes are diverged. The cover test is repeated and the procedure continued until no further movement to take up fixation is seen. The angle of deviation, in degrees, is obtained from the scale. If there is poor vision and the patient is unable to fixate with one eye, the deviation

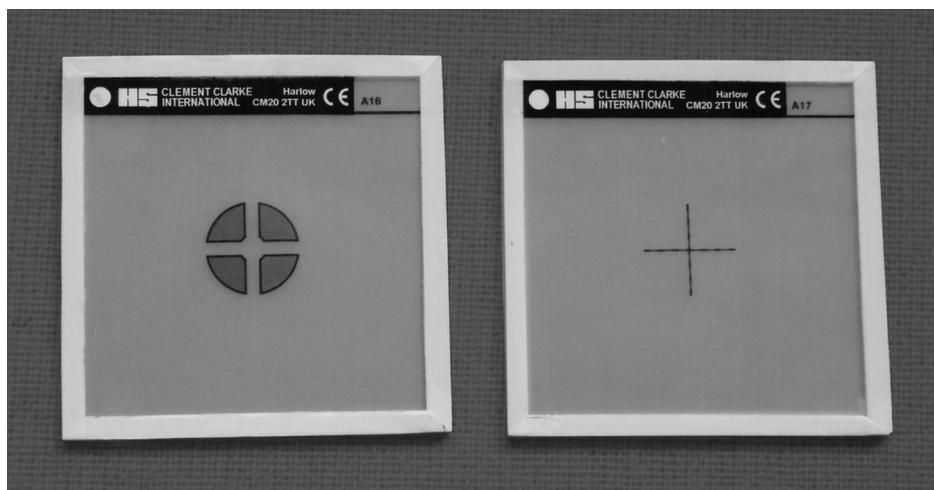


Figure 4.41 Maddox slides.

may be measured by assessing the position of corneal reflections. The tube is moved before the deviating eye until the corneal reflections are symmetrical.

When assessing the subjective angle of deviation, the patient is asked to move the tube before the deviating eye so that the images overlap. Vertical deviations are measured by moving one of the slides vertically until the patient states that there is no vertical movement of the images.

Assessment of retinal correspondence

Simultaneous perception

SP slides are dissimilar (Fig. 4.42). Subjectively, the patient moves the tube before the non-fixating eye and attempts to overlap the images. SP is absent if the patient is unable to see the two images at the same time, or if one image repeatedly disappears or the images jump. It is easier to superimpose with larger slides and if SP is not achieved with smaller slides, it is useful to reassess with peripheral slides before assuming lack of SP.

Fusion

Sensory fusion is present if, at the subjective angle, the patient is able to superimpose both fusion slide images (Fig. 4.43) and is able to see both control images (one from each slide). The motor fusion range is assessed by converging and diverging the tubes whilst asking the patient to state when fusion breaks. The patient will appreciate diplopia or will suppress one of the controls. If the patient has difficulty

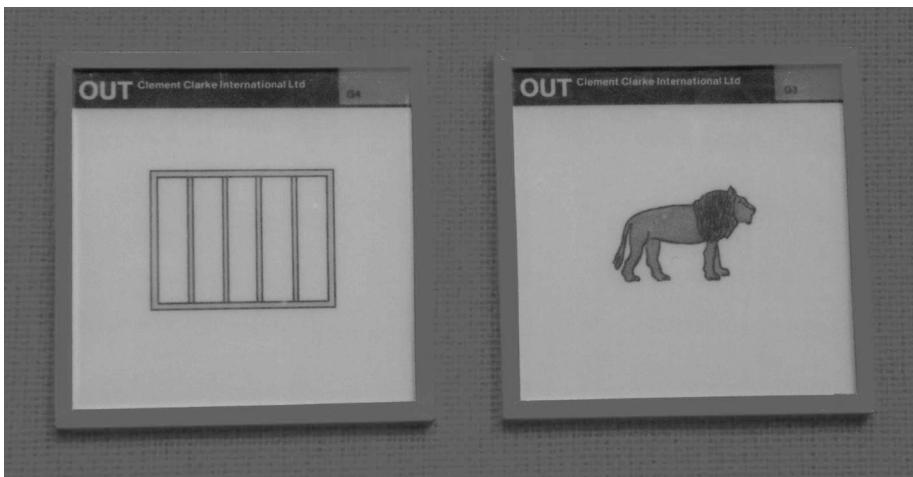


Figure 4.42 Simultaneous perception slides.

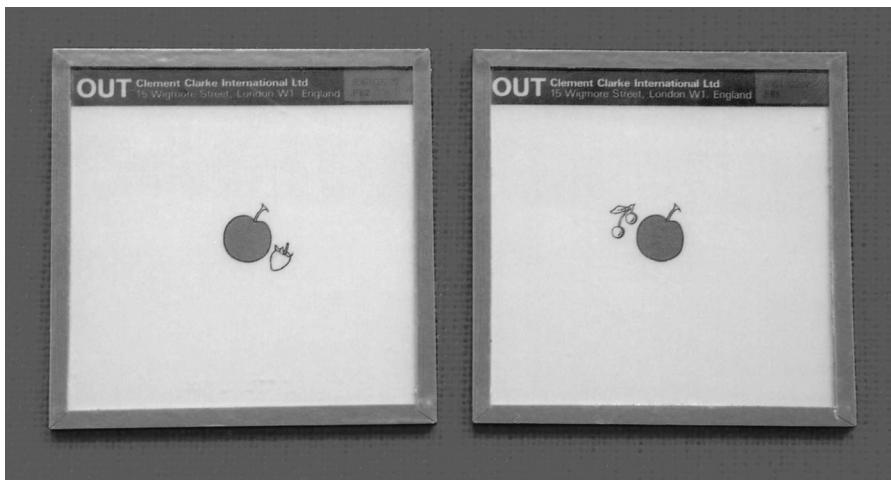


Figure 4.43 Fusion slides.

viewing the targets or loses one of the controls, larger slides may be used. Foveal fusion is more difficult to maintain than peripheral.

Stereoacuity

Both tubes are locked at the fusion angle. Gross stereopsis slides (Fig. 4.44) are inserted and the patient asked whether or not there is a stereoscopic effect. Detailed stereopsis is assessed using Braddick slides.

Assessment of aniseikonia

Ruben's slides are used to establish the difference in image size between either eye (see Chapter 9).

Assessment of after-images to investigate retinal correspondence

Refer to Section 'After-image test' for a full description.

Measurement of angle kappa

This is the angle between the pupillary axis and visual axis and is caused by a failure of the optical and visual axes of the eye to coincide. The optical axis is a line connecting the optical centres of the cornea and lens when aligned. The visual axis is the line of sight, which connects the fovea with the fixation target. Usually, the optical axis touches the retina slightly nasal and inferior to the fovea. As a result, the corneal reflection is slightly nasal to the centre of the pupil (normal positive angle kappa). If there is a large positive angle kappa, this may

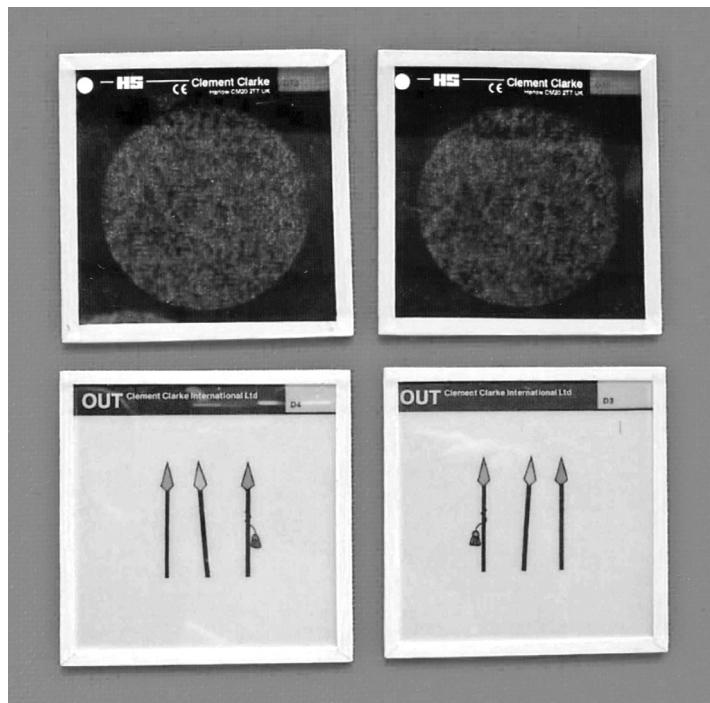


Figure 4.44 Stereopsis slides: (a) gross stereopsis; (b) detailed stereopsis.

give the appearance of a divergent deviation. If the fovea is nasal to the optical axis, the corneal reflection will appear on the temporal side of the pupil centre and this may give the appearance of a convergent deviation. Figure 4.45 illustrates angle kappa and Fig. 4.46 illustrates the slide used for measurement of the angle.

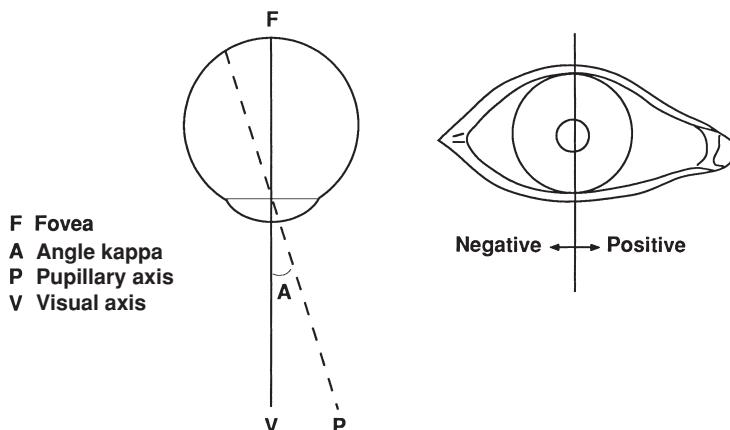


Figure 4.45 Angle kappa.

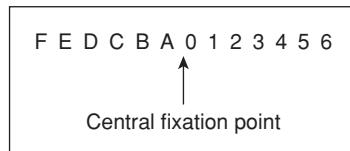


Figure 4.46 Angle kappa slide assessment.

Angle kappa is measured monocularly on the synoptophore. The slides have a horizontal series of letters and numbers separated by intervals of 1° – 2° . A row of animals is placed below the horizontal line of letters and numbers on some slides for use with children. The patient looks at the central fixation point and then on to each subsequent number until the corneal reflections appear symmetrical. This gives a measurement of a positive angle kappa. With a negative angle kappa, the patient looks at each subsequent letter progressively outwards until the corneal reflections appear symmetrical.

Haidinger's brushes

These are rarely used in current orthoptic practice but have been used in pleoptic treatment for eccentric fixation to stimulate the fovea and re-educate foveal fixation (see Chapter 8). This is an entoptic phenomenon caused by the effect of polarised light at the fovea and consists of polarised and blue filters. The background of blue light enhances this effect, which may be prolonged by rotating the axis of polarisation so that the brushes also rotate.

Aniseikonia

Space eikonometer

This is the reference test for the assessment of aniseikonia with measurement to the nearest 0.1% of image difference (Stewart & Whittle 2002).

Cover test

A qualitative assessment may be made in which the patient describes the difference in image size as the cover is moved from one eye to the other.

Brecher test

Maddox rods are held before each eye whilst the patient views two small lights. Two lights and two streaks will be seen. Any deviation is corrected and the patient is then asked to directly compare the difference between the lights and streaks.

Awaya test

Red and green goggles are worn while the patient views the Awaya test. This consists of one plate containing two equal-sized semicircles – one red and one green. If these are perceived to be the same size, aniseikonia is not present. Where a difference is seen in size, the patient then views a series of semicircles that vary in size and is asked to choose the plate that does match the control semicircle. Each semicircle changes in 1% size difference steps.

Fixation

Visuscope (fixation ophthalmoscope)

This test is used to assess fixation in cases of amblyopia and manifest strabismus. The visuscope incorporates a four-pointed star with a clear central area as the fixation target. A green filter is occasionally used to produce a red free image of the retina. The normal eye is assessed initially, to ensure patient understanding and cooperation, followed by the amblyopic eye. The position of fixation is recorded in relation to the foveola (central, parafoveal, macular, peripheral), and according to the state of fixation (steady, unsteady, wandering or eccentric) (Fig. 4.47).

Four dioptrē prism test

This test assesses the presence of bifoveal fixation. The following response is expected in the presence of normal bifoveal fixation. On placing a 4 dioptrē prism base out before the right eye, the eyes make a conjugate movement to the left. A subsequent disjunctive fusional movement of the left eye is seen. A prism placed before the left eye produces a similar fusional movement (Fig. 4.48).

In the presence of a central suppression scotoma, with the prism placed before the normal eye, a conjugate movement is seen to the opposite side. However, a

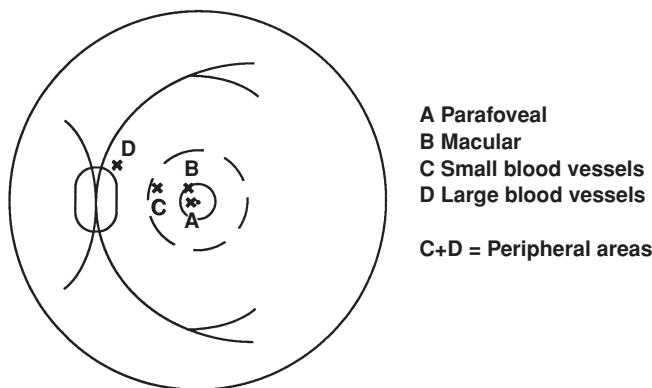


Figure 4.47 Fixation.

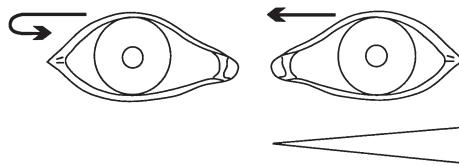


Figure 4.48 Fusion response with four dioptre prism test. When a base-out prism is placed before the left eye, both eyes move conjugately to the right. The right eye then makes a disjunctive refixation movement to the left.

disjunctive fusional movement is not seen in the affected eye because the image has been displaced into the suppression scotoma. When the prism is placed before the affected eye, no movement is seen in either eye because the image is already within the suppression scotoma (Fig. 4.49) (Bagolini *et al.* 1985).

The test may also be performed with the prism base in, particularly in cases of micro exotropia (Romano & von Noorden 1969, Epstein & Tredici 1972).

Measurement of deviations

Neutralisation

On cover test, a movement of the eyes will be observed indicating the presence of either a manifest or latent deviation. By using prisms to displace light across the retina, the deviation may be reduced and eventually neutralised when no movement of the eyes occurs. There will be no movement of the eyes when the selected prism causes the image to fall on the fovea. The prism cover test uses this principle.

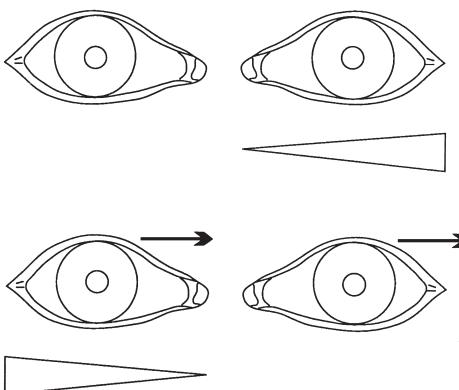


Figure 4.49 Suppression scotoma response with four dioptre prism test. When a base-out prism is placed before the left eye, no movement of the eyes is seen as the image of the fixation target falls within the left suppression scotoma. When the base-out prism is placed before the right eye, a conjugate movement of both eyes is seen to the left but there is no refixation movement of the left eye, as the image has fallen within the suppression scotoma of the left eye.

Estimation

When an accurate measurement of a deviation cannot be obtained either due to age, cooperation or poor visual acuity, an estimation of a manifest deviation may be made by observation of the corneal reflections and additionally by using a prism to displace the image until the corneal reflections appear symmetrical. The prism reflection and Krimsky tests use this method.

Objective tests

- Prism cover test
- Simultaneous prism cover test
- Prism reflection test
- Krimsky test
- Hirschberg's method
- Synoptophore

Subjective tests

- Maddox wing
- Maddox rod
- Synoptophore

Difficulties with prisms

Test accuracy is limited by the optical properties of prisms. The stronger the prism, the greater the error. When using prisms, significant errors are produced when a low power prism is added to a high power prism, that is, stacking prisms.

Errors in quantitative measurements of strabismus can also be induced by improperly placing prisms while measuring the deviation (Thompson & Guyton 1983). Glass prisms are calibrated for use in the Prentice position (posterior face of prism held perpendicular to the line of sight of the deviating eye). Plastic prisms are calibrated for use in the frontal place with the position parallel to the infraorbital rim (Fig. 4.50).

The measurement error introduced by using the prentice position increases as prisms greater than 20 prism dioptres are used. When deviations exceed the amount of the largest available prism the examiner should not stack prisms. The error can be minimised by holding one prism before each eye. However, prisms may be stacked if they are used to simultaneously measure vertical and horizontal misalignment, which are independent of each other. Prisms should be held close to the eyes as the power needed to neutralise the deviation with near fixation will increase as prisms are held further from the cornea (Thompson & Guyton 1985).

High refractive corrections may also affect measurement. Plus lenses reduce and minus lenses increase the deviation due to the prismatic effect of the spectacle correction.

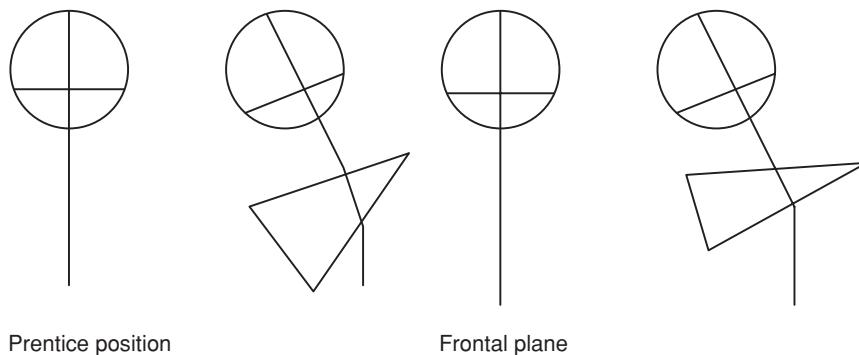


Figure 4.50 Prism position.

Prism cover test

This is an objective dissociative test used to measure the angle of deviation. The test may be performed at 1/3 m, 6 m and greater than 6 m and indicates the maximum angle of deviation. Both horizontal and vertical angles can be measured in manifest and latent deviations. The prism cover test is generally the test of choice in the majority of cases.

The patient must fixate the target accurately and have adequate visual acuity to be able to fix with either eye. The test is performed both with and without spectacles with the patient holding and fixating a target at 1/3 m, at eye level. A prism bar is held in front of the deviating eye usually in manifest deviations and before either eye in latent deviations. The other eye is occluded. An alternate cover test is performed by moving the occluder from one eye to the other whilst watching the movement of the eye behind the prism as it takes up fixation. The prism strength is then increased gradually until the movement decreases and eventually there is no movement to take up fixation. By continuing to increase the prism strength until a reverse movement is seen and then decreasing again until there is no movement, the 'null' point can be verified from both directions. The prism strength is noted and is equivalent to the maximum measurement of the angle of deviation. The test may be repeated for 6 m and greater than 6 m if required. Advantages are that the test provides accurate measurements, is performed under natural conditions and provides the maximum angle of deviation. Disadvantages are that the patient must have central fixation in either eye with at least 6/60 visual acuity to see the fixation target. It is difficult with young or uncooperative patients and prism bars have a range of only 45 prism dioptres.

Simultaneous prism cover test

The aim of this test is to neutralise the deviation without complete dissociation. It is used in cases of microtropia with an associated heterophoria, in order to isolate and

measure the angle of the manifest deviation. A prism is placed before the deviating eye and simultaneously, an occluder is placed before the other eye. The aim is to neutralise the movement of the deviating eye when it moves to take up fixation, thereby giving the angle of manifest deviation.

Prism reflection test

This test is used with small children unable to cooperate with the prism cover test or with patients with poor vision who do not have foveal fixation. The prism bar is placed before the deviating eye and the patient fixates a spotlight at 1/3 m. The examiner observes the position of the corneal reflections and the prism strength is gradually increased until the corneal reflections appear symmetrical. This prism strength is noted. The test may also be performed using loose prisms.

Krimsky test

This test is similar to the prism reflection test but the prism is placed before the fixing eye (Krimsky 1943, Rowe 1994). The patient fixates a spotlight at 1/3 m and the prism strength is increased until the corneal reflections are symmetrical. The optical principle is different to the prism reflection test as the prism is placed before the fixing eye and the fixing eye moves to overcome the prism and maintain fixation in the primary position. Because of Hering's law (each eye will make a conjugate movement of the same amount), if an increasing prism strength is placed before the fixing eye, this will centralise the corneal reflections of the deviating eye.

Hirschberg's test

This test is based on the presence of comparable corneal reflections by assessing the position of the corneal reflection in the squinting eye with that of the fixing eye.

An indication of the angle of manifest deviations can be obtained. A spotlight is shone into the patient's eyes and an estimated measurement is taken as to the angle of deviation, by observing the position of the corneal reflection of the deviating eye. One mm displacement is equivalent to 7° or 15 prism dioptres (Brodie 1987) or 1mm is equivalent to 20–22 prism dioptres in children (Hasebe *et al.* 1998). (Fig. 4.51).

Corneal reflections viewed at the pupil margin are roughly equivalent to a displacement of 10°–15° (20–30 prism dioptres), those situated half way between the pupil and the limbus, to 25° (50 prism dioptres) and those at the limbus, to 45° (90 prism dioptres). Reflections displaced temporally indicate the presence of an eso-deviation whilst nasal displacement indicates an exo-deviation.

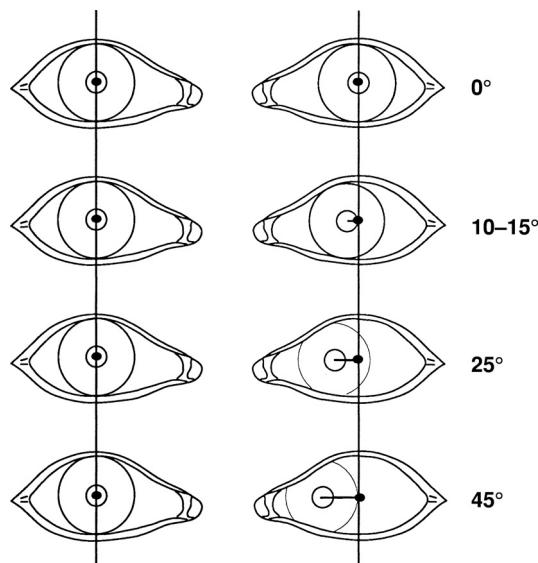


Figure 4.51 Hirschberg's corneal reflections.

Maddox rod

This is a subjective, diplopia-based test used to measure the angle of deviation in heterophorias and small heterotropias provided normal retinal correspondence is present. It is a series of parallel high power plano-convex cylinders that convert a point source of light into a line seen perpendicular to the axis of the cylinders, which are traditionally coloured red or blue. The Maddox lenses may also be in the form of a Maddox hand frame or Franceschetti frame (Fig. 4.52).

The test is performed with and without spectacles at 1/3 m and 6 m and fixating with either eye.

Horizontal deviation

The patient fixates a spotlight held at eye level. The rod is placed before the non-fixing eye and is rotated in order to produce a vertical line. The patient is asked to



Figure 4.52 Maddox rod.

state where the line is in relation to the light. If the line passes through the light, there is no horizontal deviation.

The eyes are dissociated as the stimulus to fuse has been removed because dissimilar images are presented to the eyes. If the line is seen to one side of the light, a horizontal deviation is present and the line is displaced according to the deviation present. Uncrossed displacement occurs with eso-deviations and crossed displacement with exo-deviations. In order to correct the displacement, corrective prisms of increasing strengths are placed before the rod until the patient states that the line passes through the light. This prism strength is noted.

Vertical deviation

The rod is rotated to produce a horizontal line image and the patient is asked to state the position of the line in relation to the light. If the line passes through the light, there is no vertical deviation. If the line is above the light, there is a hypo-deviation, and if below the light, there is a hyper-deviation. Corrective prisms of increasing strengths are used until the patient states that the line passes through the light. This prism strength is noted.

Maddox wing

This is a subjective test used to measure the angle of deviation in heterophorias and small heterotropias provided normal retinal correspondence is present. The instrument consists of a metal plate bearing a horizontal row of white numbers graded from right to left (22...01...15) and a white arrow. It also contains a vertical row of numbers graded from top to bottom (22...01...13), and a red arrow.

The wing is situated at 1/3 m from the eyes and is supported by a septum and handle, which connects it to the eye pieces containing trial lens holders. The septum is arranged so that the patient sees the arrows with the right eye and numbers with the left eye. The test is performed with and without spectacles and the patient asked to state which white number the white arrow is pointing to. If it points to an even number, there is a divergent deviation and an odd number is indicative of an eso-deviation. The size of deviation corresponds in prism dioptres to the number stated.

For vertical deviations, the patient observes the red arrow and states which red number it points to. Between the numbers 22...01, there is a left/right deviation and between 01...13, there is a right/left deviation.

Torsion may also be assessed by asking the patient whether the red arrow is parallel to the white line of numbers. If not, the arrow can be moved until it is parallel and the amount of torsion recorded in degrees from the scale situated at the right side of the metal plate (Fig. 4.53).

Synoptophore

The uses of this instrument are described in detail earlier in the chapter.

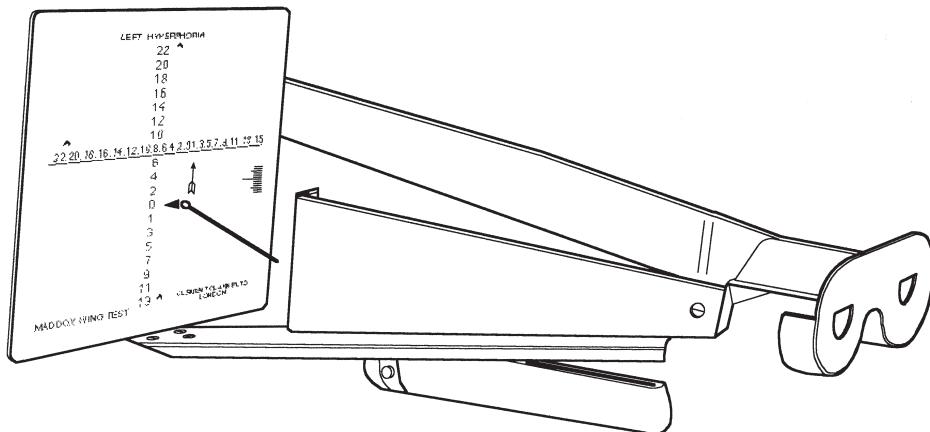


Figure 4.53 Maddox wing.

Hess charts

Hess charts are used in the investigation of incomitant strabismus in order to assess the paretic element (see Chapters 14–16). By means of dissociation, it is possible to indicate the position of the non-fixing eye when the other eye is fixing, in specified positions of gaze. A field is plotted in this way for each eye. Each point on the inner field represents fixation at 15° from the primary position and on the outer field, at 30° . Defects that are present on more extreme gaze than this will not necessarily be evident on the Hess chart. It is essential to interpret these results together with the assessment of ocular movements. Information is gained from the chart by comparing the fields with each other and with the normal field printed on the chart.

Hess screen

This consists of a screen incorporating a tangent pattern on a black/grey background (Fig. 4.54). It may be manually or electrically operated. Red and green glasses are worn by the patient, with the red filter before the fixing eye. The patient is seated at 50 cm from the screen.

Dissociation is achieved by means of complementary colours. The fixing eye sees only a tangent pattern with red points or lights while the deviating eye sees only a green spot projected into the screen by a torch held by the patient. There must be macular/macular projection to ensure that either eye can maintain fixation. Therefore, the position of the field will reflect the true extent of the deviation.

Lees screen

This consists of two opalescent screens placed at 90° to each other and a two-sided mirror septum at 45° to the screens, which dissociates the visual fields (Fig. 4.55).

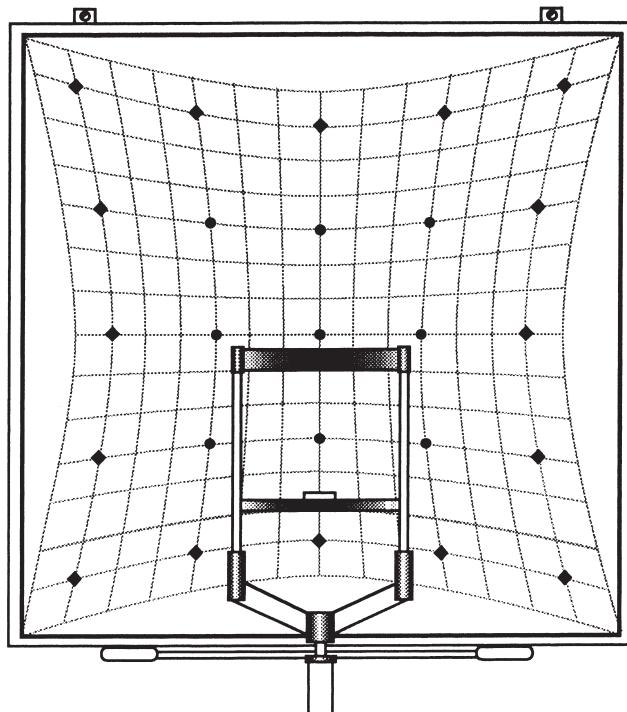


Figure 4.54 Hess screen.

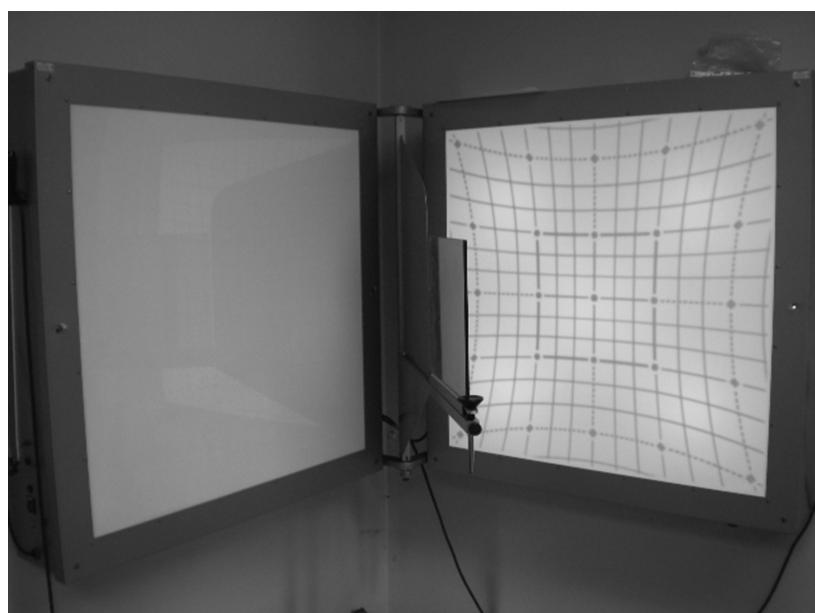


Figure 4.55 Lees screen.

The mirror must be a true plane mirror and bisect the screens exactly. Each screen incorporates a tangent grid with an inner and outer field. There must be macular/macular projection.

The Lees screen possesses a number of advantages over the Hess screen:

- Accurate dissociation
- Greater contrast between tangent scale and background
- Increased ease and speed of performance
- Constant illumination

Interpretation of results

The patient must possess normal retinal correspondence and central fixation. The test may be performed in the presence of abnormal retinal correspondence if a record of motility is required for comparison from visit to visit.

Comparing fields with each other

1. *Size of fields*: The smaller field represents the affected eye. Any difference in size indicates incomitance, which is usually indicative of recent onset paresis. Similar size indicates concomitance.
2. Sloping sides to fields indicates an A/V pattern.

Comparing each field with the normal field

1. *Smaller field*: The deviation in the primary position is the primary deviation. The greatest negative (inwards) displacement represents the primary underaction and is seen in the direction of the main action of the affected muscle. Positive (outwards) displacement indicates an overaction.
2. *Larger field*: The deviation in primary position is the secondary deviation. The greatest positive displacement is the main overaction and the maximum displacement often occurs in the direction of the main action of the overacting contralateral synergist in the larger field. It is important to note any difference in the deviation in other positions of gaze.

Outer fields

These may show a defect when the central fields appear normal, particularly with mechanical defects or very slight paresis.

Information obtained

1. Underactions and overactions.
2. Affected eye.
3. A or V pattern.

4. Type of deviation in the primary position and other positions of gaze.
5. Size of deviation. Each box on the chart represents 5°.
6. Primary and secondary deviations.
7. Muscle sequelae. There are limited sequelae with mechanical defects. The presence of full or part developed muscle sequelae aids in the differential diagnosis of concomitance and incomitance and recent and long-standing deviations.
8. Comparison between clinic visits.
9. Comparison between pre- and post-operative visits.
10. It is possible to measure torsion using the Lees screen. This is described later in the chapter.

Characteristics of mechanical defects

1. Compressed fields.
2. Limited muscle sequelae.
3. The deviation in the primary position does not reflect the extent of the defect. Binocular single vision is often present in the primary position.

Characteristics of neurogenic defects

1. Sloping fields. These are indicative of an A or V pattern, not torsion.
2. Muscle sequelae are not limited and will eventually attain relative concomitance.
3. The deviation in the primary position reflects the extent of the palsy and therefore binocular single vision may or may not be present in the primary position.

Field of binocular single vision

The binocular field is plotted using a perimeter (Arc, Goldmann or Octopus) (Figs. 4.56–4.58) with a single chin rest using a letter or III4e stimulus (Shotton *et al.* 2008, Rowe & Hanif 2011). The single chin rest is used to place the head in a central position with both eyes level and aligned with the fixation target. It is plotted for patients with incomitant deviations who appreciate diplopia but have an area of single vision and is used to monitor the progress of the condition, the effects of any treatment and for pre- and post-operative comparison. The patient is seated level with the fixation target and with the eyes in the primary position.

With both eyes open, the patient observes the target as it is moved slowly away from the primary position and is asked to state when it appears double. At this point, the examiner stops and marks the chart. The target is returned to the primary position and the arc is rotated (usually through 30° intervals) and the test repeated through 360°. On completion, the marked dots on the chart are joined and the area of single vision is shaded.



Figure 4.56 Arc perimeter.



Figure 4.57 Goldmann perimeter.



Figure 4.58 Octopus perimeter.

At certain points, the facial contours (e.g. the nose) may obscure one eye. A cover test should be performed to ensure single vision is present because the patient will not appreciate diplopia in this situation. The test is performed without spectacles because the frame obscures vision.

If diplopia is present in the primary position, the target is moved into the area of single vision, recorded and moved again until diplopia is again perceived.

The field of binocular single vision can be scored using various measurement templates (Woodruff *et al.* 1987, Fitzsimons & White 1990, Sullivan *et al.* 1992, Shotton *et al.* 2008). Most are weighted with increased scores assigned to primary and depressed gaze.

Unicocular field of vision

This may be plotted using an arc perimeter with a double chin rest, which places the head in an off centred position and each eye aligned with the fixation target in turn. One eye is occluded during the test and the patient must maintain foveal fixation, which is monitored by ensuring that the patient perceives clear vision. The test is indicated in patients who have very limited ocular movements and where responses obtained from the Hess chart are confused and overlapping.

Fixation target is typically a small letter that blurs when not fixated foveally. A red target may be used as an alternative. The test can also be plotted using a Goldmann or Octopus perimeter and the foveal threshold value can be determined. Adaptations have considerably improved test/retest and intraobserver reliability by measuring the field using only six positions (Fig. 4.59) corresponding to the main action of each extraocular muscle (Haggerty *et al.* 2005, Hanif *et al.* 2008, Rowe & Hanif 2011).

Right eye:
(left eye mirror image)

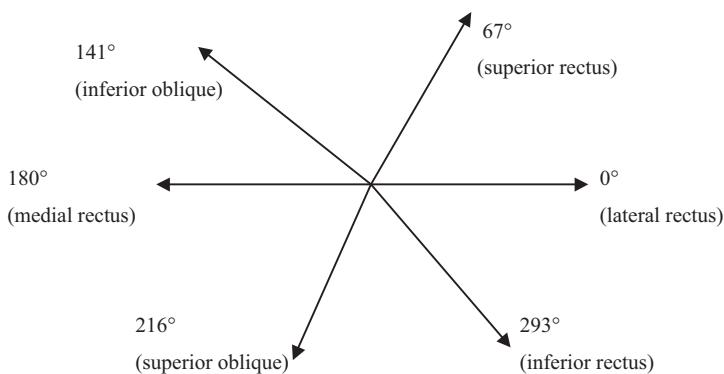


Figure 4.59 Six vectors for uniocular rotations.

Measurement of torsion

Torsion may be measured objectively or subjectively. An objective measure is obtained by observing the fovea-optic disc relationship ophthalmoscopically in congenital cases. If torsion is present, the disc will appear displaced and either intorted or extorted in relation to the fovea (Fig. 4.60) (Bixenman & von Noorden 1982). Subjective measurements include the following:

- Synoptophore
- Adapted Lees screen
- Maddox wing
- Awaya cyclo test
- Bagolini glasses
- Maddox double rod

Synoptophore

Maddox slides are used and the degree of torsion is measured subjectively. The cross is placed before the fixing eye and the segmented circle is placed before the non-fixing eye. The patient superimposes the cross within the segmented circle, so that all lines are parallel at 90° and 180°. Torsion is adjusted by rotating the slide with the tilted image and the amount recorded in degrees. Torsion may be assessed fixing either eye and in the nine cardinal positions of gaze.

Adapted Lees screen

Linear targets are used (Dulley & Harden 1974). The patient places a short bar, mounted on a pointer, on sequential dots with the bar lying along the horizontal

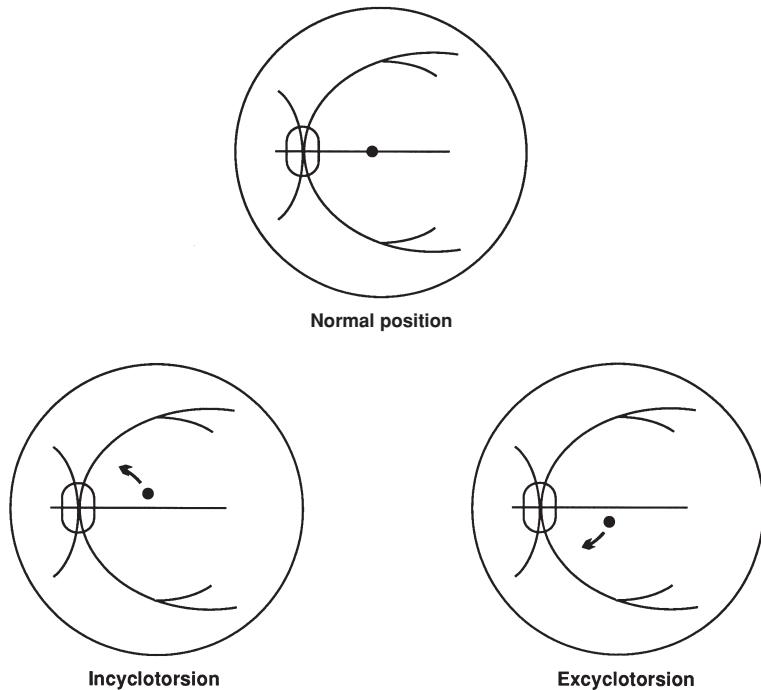


Figure 4.60 Objective assessment of torsion.

grid line. If torsion is present, the bar will slope and the position of its ends should be marked on the screen using a non-permanent marker. The amount of torsion is measured on completion of the test, using a protractor mounted on a Perspex block, which is pivoted around a fixed centre of rotation. The block is placed parallel to the horizontal grid line and the protractor is rotated until its base is in line with the marked position of the bar. Measurement of torsion is given in degrees.

Maddox wing

Torsion is present if the horizontal red arrow is not parallel to the white line composed of numbers. The arrow may be moved until it is parallel and the amount of torsion is indicated on a separate measurement scale. Each division on the scale is equivalent to 2° of arc. The arrow is usually moved by the patient giving a subjective measurement.

Awaya cyclo test

The test is presented in book form presenting 13 pairs of half moons. One half of each pair is red and the other green and each pair are spaced 10 mm apart. The green half moons are tilted in gradations of 1° . Initially, the plate of parallel half moons is shown to the patient. Should the green half moon appear tilted clockwise, there is extorsion and if it appears tilted anticlockwise, there is intorsion. The

patient wears red and green glasses with the red filter before the right eye if there is extorsion and the red filter before the left eye if there is intorsion. The patient decides which pair of half moons are parallel and this measurement is noted.

Bagolini glasses

Two Bagolini glasses are placed in a trial frame, one before either eye. The striations are placed vertically so that two horizontal line images are seen. In the presence of a vertical deviation, the lines will be seen one above the other. If there is no vertical deviation, a small vertical prism is introduced in order to separate the images. The patient is asked to state whether or not the lines are parallel. If tilted, the image can be straightened by rotating one of the Bagolini glasses and the degree of intorsion or extorsion measured from the trial frame. The test may be performed with either eye fixating (Ruttmann & von Noorden 1984).

Maddox double rod

The test is conducted in the same manner as that described above for Bagolini glasses. Two Maddox rods are placed in a trial frame with the cylinder axes vertical. Large variations are noted with large degrees of torsion (Freedman *et al.* 2003). Thus, results should be treated with caution.

Parks-Helveston three-step test

This triple procedure may assist in the diagnosis of vertical muscle palsies:

1. A cover test is performed in the primary position and the hypertropic eye is recorded.
2. An alternate cover test is then performed on dextroversion and laevoversion and the direction of the greater vertical deviation is recorded.
3. The Bielschowsky head tilt test is performed. This differentiates between a superior oblique and a contralateral superior rectus palsy.

The patient fixates a target at 3 m and a cover test is performed in the primary position and with the patient's head tilted maximally to the left and then to the right. The difference in the degree of vertical deviation is recorded. An increase in the vertical angle when the head is tilted to the affected side is indicative of a superior oblique palsy. However, results can be misleading with regard to differential diagnosis. A positive test results has been observed with superior rectus palsy, inferior oblique overaction and cases of DVD.

Diplopia charts

These are used to evaluate and document the type of diplopia present. Red and green glasses are worn with the red filter usually placed before the right eye. A linear light source is used so that torsion may also be assessed. The patient's

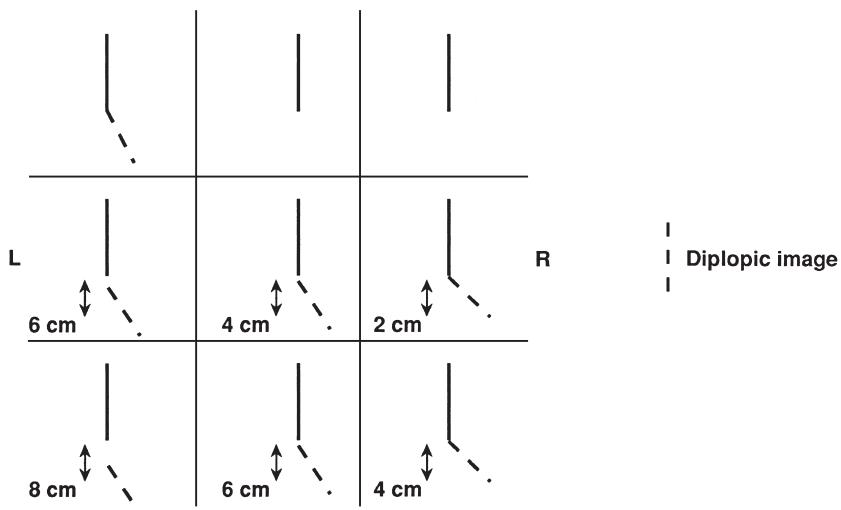


Figure 4.61 Diplopia chart of IV nerve palsy.

head should be still and erect. The patient is asked to describe diplopia in the nine cardinal positions of gaze starting with the primary position and also to describe the relative position of the lines of light in each position of gaze. The results are recorded in chart form as if one is looking through the patient's eyes. The position at which diplopia occurs, the type of diplopia, the maximum separation of images and the distal image are all recorded (Figs. 4.61 and 4.62).

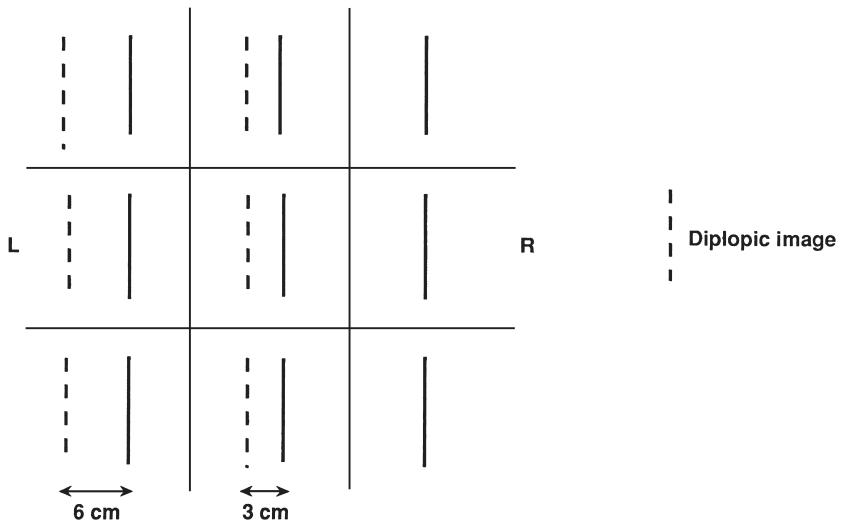


Figure 4.62 Diplopia chart of VI nerve palsy.

Bielchowsky phenomenon (dark wedge test)

This is a clinical test used to differentiate between DVD and alternating hyperphoria. One eye is occluded (a Spielmann occluder is preferable) and is seen to elevate under cover. With a spotlight at 6 m, a Sbisa bar (Fig. 4.37) is placed before the fixating eye and the filter density is gradually increased. The examiner observes the occluded eye that will make a downwards movement and may often pass below the midline, if DVD is present. The occluded eye will start to elevate when the filter density is again decreased. By contrast, the elevated position of the occluded eye will remain unchanged when increasingly dense filters are placed before the fixing eye in cases of alternating hyperphoria.

Forced duction test

This test is used to assess the passive movement of the globe when active movement is restricted. It is used to determine the tone of the muscle by attempting to move the eye with forceps in different directions. The eye is anaesthetised and then gripped at opposite limbal points with two pairs of fine-toothed forceps. The eye is rotated into each position of gaze to assess the extent and ease of eye movement. A limitation of passive movement indicates a mechanical restriction (Stephens & Reinecke 1967).

Forced generation test

This test assesses the active muscle force in eye movement. By evaluating the amount of contraction force present in an apparently paretic muscle, an indication of muscle potential is gained. The anaesthetised eye is gripped at opposite limbal points with two pairs of fine-toothed forceps and held firmly. The patient is asked to move the eyes in the direction of gaze of the limited movement. The extent of muscle action can be felt by the examiner during the attempt to rotate the eye.

Orthoptic exercises

Orthoptic exercises are indicated in certain conditions:

- To increase control in latent deviations
- To increase control in intermittent deviations
- Pre- or post-operative to consolidate the result
- To achieve good binocular convergence

Patients must be well motivated before receiving exercises.

Bar reading

Bar reading is controlled reading that allows accommodation without overconvergence. It is based on the principle of crossed physiological diplopia so that when

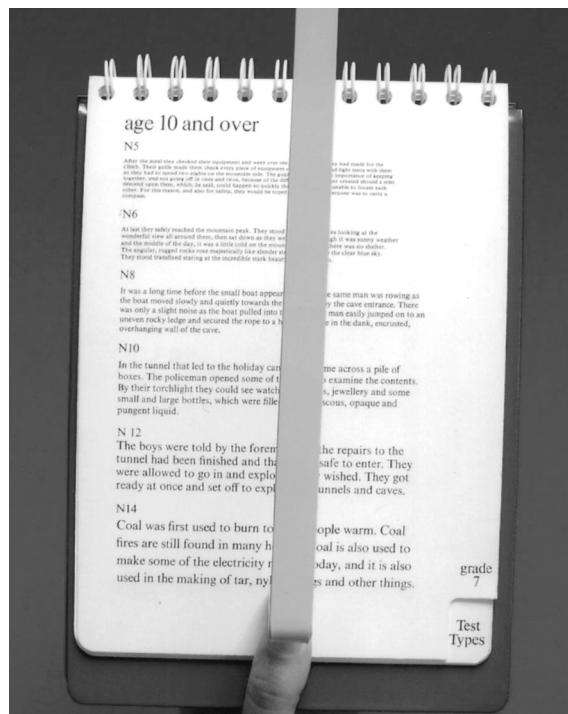


Figure 4.63 Bar reading.

the subject looks at the print, the bar in front of the print is seen as a blurred double image. If the subject is binocular, the subject will be able to read as one eye will always see the text. If not binocular, the bar will hide part of the text. The patient's binocular visual acuity is assessed and the test commenced with print that is one size larger than the level of binocular visual acuity. The patient holds the book with the bar held against the bottom of the page. Use of high contrast bars may make the task easier as there is greater contrast of the bar against the text. However, no difference has been shown for high or low contrast coloured bars in normal subjects (Rowe & Tremarco 2010). The bar will thus be at 8 cm from the page. The bar must be held steadily with the book at 1/3 m in a slightly depressed reading position. The patient is then instructed to read along the lines of the book (Fig. 4.63).

The head must be kept still and erect so that the patient does not look around the bar. Where binocular single vision is present, when the bar obstructs the vision of one eye looking at a word, the word is still seen by the other unobstructed eye and reading is therefore not impaired by the presence of the bar.

Indications for use

- Treatment of intermittent fully accommodative esotropia.
- To improve binocular single vision.

- Treatment of decompensating heterophoria.
- Differential diagnosis between intermittent fully accommodative esotropia and convergence excess esotropia. Bar reading is not achieved with cases of convergence excess esotropia.

Stereograms

Stereograms are based on the principle of physiological diplopia, the ability to fuse dissimilar images giving rise to stereoscopic vision, and relative convergence.

Types of stereograms (Fig. 4.64)

- Non-stereoscopic – cats.
- Stereoscopic – buckets (clear and filled), RAF (ABC or 123).

The patient must have some form of binocular single vision to be able to appreciate stereograms.

Stereograms may be used in two positions using either near or distance fixation.

Near position

This position induces positive relative accommodative convergence. When the stereoscopic effect is perceived, the patient's eyes are converged on the fixation target but accommodation is exerted to a lesser extent to perceive a clear image of the images on the stereogram card.

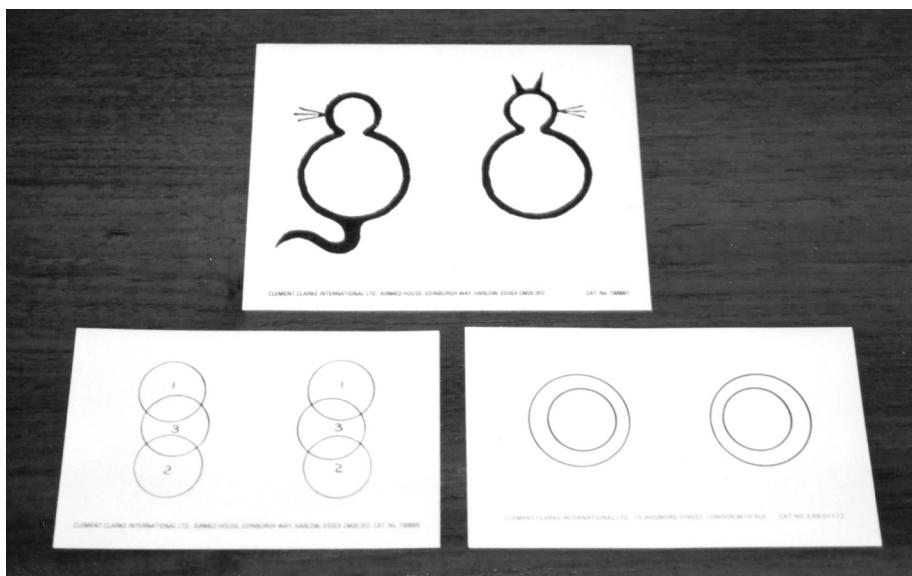


Figure 4.64 Stereograms.

The card should be held at arms length in the primary position and the patient fixates a target between the card and the eyes. Three images should be appreciated on the card. If four images are seen, the position of the target should be adjusted until the middle two images are fused. The middle image should have all control features. When using a stereoscopic stereogram, ensure that depth is perceived. The patient should aim to see the three images clearly.

Distance position

This position induces negative relative convergence. When the stereoscopic effect is perceived, the patient's eyes are aligned on the distant fixation target but accommodation is exerted to a greater extent to perceive a clear image of the images on the stereogram card.

The patient fixates a target in the distance in the primary position and the card is held at about 1/3 m from the nose. The upper border of the card should appear to be just underneath the distance target. Three images should be appreciated and the middle image should have all control features present. The patient should aim to see all three images clearly.

Indications for use

- Cases of symptom producing heterophoria. Stereograms are used in the near position with divergent deviations and in the distance position with convergent deviations.
- Convergence insufficiency.
- Intermittent fully accommodative esotropia.
- Intermittent exotropia.

Methods to overcome suppression

1. *Occlusion or monocular reading:* This may be used in conjunction with amblyopia treatment. The fixing eye is occluded so that the deviating eye takes up fixation. This is combined with hand/eye coordination exercises.
2. *Red filter drawing:* The patient wears a red filter over the deviating eye and an occluder over the other eye. A complementary coloured pen is used. This exercise may be used for 10–15 minutes daily. Diplopia is appreciated when the filter and occlusion is removed.
3. *Recognition of diplopia:* This is carried out with red and green glasses and a spotlight.
4. *Sbisa bar:* This is placed before fixing eye in increasing density until the deviating eye fixates and diplopia is appreciated. The filter density is gradually reduced so that the deviating eye is still used and diplopia present continually.

5. *Synoptophore*: Suppression exercises include chasing (examiner moves one tube and patient chases with the other tube), flashing the light in the tube before the deviating eye, kinetic stimulation, tapping (moving the slide image before the deviating eye), and altering the rheostat intensity. Mayou slides may be used where the patient places a target into large boxes and the box size is gradually decreased.
6. *Septum*: This dissociates the eyes in free space using an opaque card between the eyes.
7. *Vertical prism*: This overcomes vertical suppression. The prism strength is gradually decreased while diplopia is appreciated.
8. *Cheiroscope*: The patient looks through two eye pieces that are separated by an oblique mirror septum that is placed at an angle of 45°. One eye sees a pad of paper and the other eye sees a pen that the patient is holding. The patient is instructed to draw pictures/shapes.

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SECTION II

5 Heterophoria

Where heterophoria is present, both visual axes are directed towards the fixation point but deviate on dissociation. Orthophoria occurs where both visual axes are directed towards the fixation point and do not deviate on dissociation.

Classification

- Esophoria
 - Convergence excess
 - Divergence weakness
 - Non-specific
- Exophoria
 - Convergence weakness
 - Divergence excess
 - Non-specific
- Hyperphoria/hypophoria
- Alternating hyperphoria/hypophoria
- Cyclophoria

Aetiology

Esophoria

- *Anatomical:* Enophthalmos, narrow IPD, extraocular muscle anomalies
- *Refractive:* Suprable hypermetropia, anisohypermetropia
- High AC/A ratio
- Weak negative fusional reserves

Exophoria

- *Anatomical:* Exophthalmos, wide IPD, extraocular muscle anomalies
- *Refractive:* Acquired myopia, presbyopia, anisomyopia

- Weak positive fusional reserves
- Passage of time

Hyperphoria/hypophoria

- *Refractive:* High myopia (heavy eye syndrome)
- Weak vertical fusional reserves
- *Anatomical:* Displaced globes, abnormal extraocular muscles, ptosis

Cyclophoria

- Oblique astigmatism

A compensated or controlled heterophoria is asymptomatic and accounts for the majority of the population. Decompensation occurs with the inability to control the angle of deviation through inadequate fusional amplitudes, which produces symptoms in older age groups. The symptoms are due to the failure to maintain binocular single vision or due to the effort required to maintain binocular single vision.

Causes of decompensation

- *Optical:* Under-, over- or uncorrected refractive errors. Wrongly prescribed spectacles. Ill-fitting spectacles. Aniseikonia.
- *Medical:* Poor general health. Fatigue. Head injury affecting fusional abilities. Drugs affecting accommodation. Alcohol. Dissociative/monocular viewing. For example, an eye pad for an ocular infection may decompensate an existing large-angle heterophoria. Visual field loss.

Esophoria

The eyes become convergent on dissociation.

- *Convergence excess:* Esophoria is greater on near fixation than distant fixation.
- *Divergence weakness:* Esophoria is greater on distant fixation than near fixation.
- *Non-specific:* Esophoria that does not vary significantly in degree for any distance.

Exophoria

The eyes become divergent on dissociation.

- *Convergence weakness:* Exophoria is greater on near fixation than distant fixation.
- *Divergence excess:* Exophoria is greater on distant fixation than near fixation.
- *Non-specific:* Exophoria that does not vary significantly in degree for any distance.

Hyperphoria/hypophoria

A vertical deviation occurring on dissociation in which one eye rotates upwards and the other downwards depending on the fixation.

Alternating hyperphoria

Either eye rotates upwards on dissociation as a result of extraocular muscle imbalance.

Alternating hypophoria

Either eye rotates downwards on dissociation as a result of extraocular muscle imbalance.

Cyclophoria

Either eye wheel rotates on dissociation, so that the upper end of the vertical axis is nasal (incyclophoria) or temporal (excyclophoria).

Incomitant heterophoria

The angle of deviation differs fixing either eye and in different positions of gaze. This is often due to an extraocular muscle underaction and treatment usually involves use of prisms or surgery.

Hemifield slide

Significant visual field loss in either eye that includes the macula may result in loss of fusion and dissociation of heterophoria to esotropia or exotropia. Complete bitemporal deficit in particular interferes with fusion. The loss of temporal visual field in both eyes disrupts stimulation of corresponding retinal points and results in failure to generate corrective fusional vergences leading to decompensation of heterophoria (Fisher *et al.* 1968, Roper-Hall 1976, Fritz & Brodsky 1992).

Hemifield slide occurs where the visual fields of the two eyes are separated from each other as in bitemporal hemianopia and cannot be joined by the normal fusional vergence reflex, creating reading problems (Shainberg *et al.* 1995, Rowe 1996). Patients have a blind area distal to the fixation point, which is termed post-fixational blindness (Fig. 5.1). When they fixate bifoveally, they are unable to locate targets beyond the fixation target and cannot respond to uncrossed physiological diplopia or base-in prisms because of the loss of divergence disparity (Shainberg *et al.* 1995, Rowe 1996).

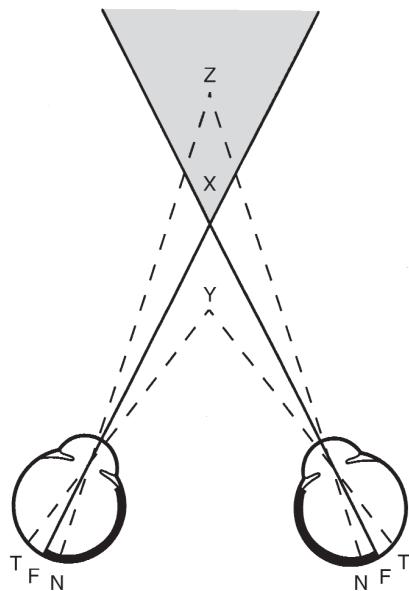


Figure 5.1 Post-fixational blindness. F is the fovea in each eye. X is the object of fixation and Z is a point behind the point of fixation. The image falls on impaired nasal retina (N) and is not seen. Y is a point in front of the point of fixation. The image falls on temporal retina (T) and is seen as physiological crossed diplopia.

Investigation of heterophoria

Case history

Symptoms of decompensation are those of headaches, asthenopia, blurring of print, intermittent diplopia and confusion if the deviation has become manifest. Pre-disposing factors include a change in occupation, poor general health, general fatigue and change in refractive error (Muhlendyck 1979).

Visual acuity

Ensure acuities are appropriately corrected with any required refractive error.

Cover test

The rate of recovery should be noted and the size of heterophoria as these factors will denote the state of control of the deviation.

Convergence

This is important with exo-deviations as a reduced or remote near point indicates decompensation, in part, of the exophoria.

Binocular function

Fusional reserves may be reduced and it is necessary to assess the range of fusional convergence and divergence that are used to compensate for exo- and eso-deviations, respectively. The order of fusion testing should be altered with base-out range tested first for exophoria and base-in range first for esophoria (Antona *et al.* 2008, Rosenfield *et al.* 1995, Antona *et al.* 2008, Rowe 2010). This allows determination of maximum opposing vergence range for control of the deviation. Binocular visual acuity and controlled binocular acuity will show decompensation when accommodation is interrupted.

Measurements The near point of accommodation, stereopsis and angle of deviation are assessed.

Monocular diagnostic occlusion Ocular and non-ocular symptoms may be differentiated by using monocular diagnostic occlusion during reading for a period of at least half an hour. Symptoms will improve if ocular.

Management

Indications for treatment include the following:

- Symptom-producing deviations
- Children where there is a possibility of decompensation in later life
- Circumstances, where necessary to achieve specific standards required for certain careers

Aims of treatment include the following:

- Reducing the deviation to within the fusional amplitude
- Improving fusional vergence
- Improving fusional amplitude

Refractive error

Correct any existing refractive error as an uncorrected or undercorrected refractive error may result in decompensation of a heterophoria. Generally, hypermetropia ≥ 1.50 dioptres, myopia ≥ 1.00 dioptres, astigmatism ≥ 1.00 dioptres and anisometropia >1.00 dioptres difference, should be corrected. Progressive addition lenses have been found to slow the progression of myopia compared to single-vision lenses in children with high accommodative lag and near esophoria (COMET 2 PEDIG 2011).

Orthoptic exercises

For esophoria, it is important to improve negative relative convergence with stereograms using distance fixation, bar reading and fusional exercises with base-in prisms and the divergent range on the synoptophore (Bryer 1961, Salah *et al.* 2000). For exophoria, it is important to improve positive relative convergence using near fixation, binocular convergence, fusional exercises with base-out prisms and the convergent range on the synoptophore (Neikter & Eriksson-Deronet 2004).

Prisms

They may be used with moderate- or large-angle deviations. The deviation should be only partially corrected (often half the prism cover test measurement) so that fusional vergences continue to be exercised (Paris & Weiss 1995). Prisms may be

incorporated into the spectacle correction for long-term management (O'Leary & Evans 2006).

Surgery

With moderate or large angles it may be necessary to reduce the angle surgically as fusional vergences may not be sufficient to maintain comfortable control at all times (Neikter *et al.* 2004).

Botulinum toxin

This can be used both diagnostically and therapeutically and a single administration may be sufficient to allow a return to asymptomatic binocular single vision. Injection to the medial rectus is undertaken for esophoria and to the lateral rectus for exophoria. Asymptomatic outcome or improved control is achieved in roughly 50% (Ripley & Rowe 2007, Bansal *et al.* 2008).

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Further reading

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6 Heterotropia

Heterotropia is a condition in which one or other visual axis is not directed towards the fixation point. The incidence of strabismus is 5–8% in the general population. One population-based study reports esotropia as more common than exotropia with strabismus being significantly associated with prematurity, visual impairment, refractive error and amblyopia (Robaei *et al.* 2006). A further population-based study reported that the incidence rate for esotropia was greatest in the first 5 years of life with fully accommodative and acquired non-accommodative esotropia types being most common (Greenberg *et al.* 2007).

Concomitant strabismus is strabismus in which the angle of deviation remains the same in all directions of gaze, whichever eye is fixing.

Esotropia

Esotropia is a condition in which one or the other eye deviates nasally.

Classification

See Fig. 6.1.

Aetiology

Primary exotropia

Constant

- *Non-accommodative:* Anatomical (narrow inter-pupillary distance [IPD], enophthalmos, muscle anomalies), proprioceptive, hereditary, congenital nystagmus, high congenital myopia, unknown
- *Accommodative:* Anatomical, proprioceptive, hereditary, always with hypermetropia or high AC/A ratio

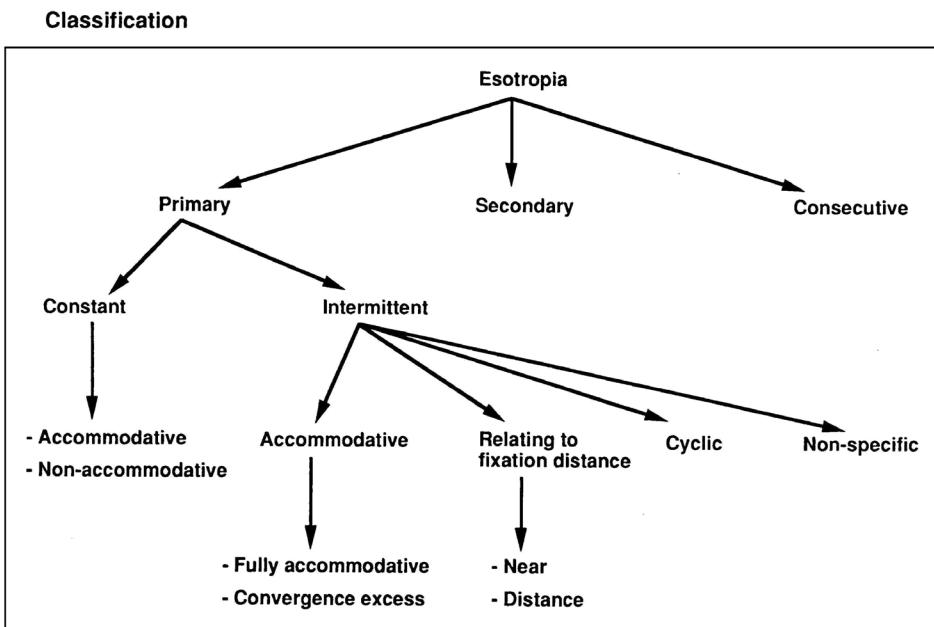


Figure 6.1 Classification of esotropia.

Intermittent

- *Fully accommodative:* Suprable hypermetropia, < +6.0 DS
- *Convergence excess:* High AC/A ratio
- *Near:* Proximal convergence, muscle anomalies
- *Distance:* Muscle anomalies
- *Cyclic:* Biorhythms, general hyperactivity of the central nervous system
- *Non-specific:* Any of the above, weak fusion

Secondary exotropia

- *Loss/impairment of vision:* Six months to 7 years as accommodative convergence is most active

Consecutive exotropia

- Surgical overcorrection of divergent deviation

Factors necessary for development of binocular single vision

1. Normal anatomy of the eye and orbit
2. Normal refractive systems

3. Normal sensory, psychomotor and motor pathways
4. Normal mental development

Interruption of any of these factors can result in development of a manifest deviation.

Primary esotropia

The convergent deviation constitutes the initial defect.

Constant esotropia with an accommodative element

The esotropia is present under all conditions and increases when accommodation is exerted.

Investigation

<i>Case history</i>	An early, insidious onset, generally between 1 and 3 years is documented. Check for precipitating causes and family history.
<i>Refraction</i>	A hypermetropic error, often associated with astigmatism and anisometropia, is detected.
<i>Visual acuity</i>	Amblyopia may be present, particularly in cases of constant unilateral strabismus.
<i>Cover test</i>	The angle of deviation decreases when hypermetropia is corrected and may be less marked for distance fixation, although the deviation remains manifest. There may be an associated dissociated vertical deviation (DVD) (see later in Chapter 6) and/or vertical deviation that is most often due to inferior oblique overaction.
<i>Convergence</i>	Convergence to 6 cm with the same angle of deviation indicates abnormal retinal correspondence (ARC) (Moore 1990). Convergence with reducing angle of deviation to intersection of the visual axes followed by symmetrical convergence of either eye indicates normal retinal correspondence. Divergence following intersection of the visual axes indicates absence of retinal correspondence. Retinal correspondence cannot be estimated where there is convergence to 6 cm with reducing angle of deviation as intersection of the visual axes does not occur.
<i>Binocular function</i>	Free space tests may be used to assess the presence of ARC with the strabismic angle of deviation and normal retinal correspondence at the corrected angle. A binocular response is indicative of ARC, but will generally be seen in small angle deviations. Free space fusion and stereopsis may be further tested to examine the quality of ARC. Suppression responses on free space tests do not give an indication of retinal correspondence.

Retinal correspondence may be evaluated in full using the synoptophore. Normal retinal correspondence is present where superimposition is achieved at the full corrected angle of deviation. ARC is present where superimposition is achieved at the angle of deviation. The quality of retinal correspondence, whether normal or abnormal, may be determined by assessing the fusion and stereopsis responses at these angles.

Where suppression responses are obtained, it is not possible to determine retinal correspondence. Therefore, further assessments may be undertaken including afterimages and using vertical prisms to move the image out of the suppression area and align the vertically displaced images using horizontal prisms. Where there is suppression and the patient is aged 7 years or greater, the depth and area of suppression should be measured in order to assess the risk of post-operative diplopia.

<i>Angle of deviation</i>	This should be measured with and without the hypermetropic correction and for near and distance fixation.
<i>AC/A ratio</i>	This is assessed preferably by the gradient method and may be high.

Management

Correct the refractive error and treat amblyopia if present. The angle of deviation often reduces following amblyopia treatment resulting in fewer cases that actually require surgical management for the strabismus (Koc *et al.* 2006). Surgery may be for a corrective or functional result.

- *Corrective:* The aim is to undercorrect the deviation to guard against a later onset of consecutive exotropia.
- *Functional:* The aim is to obtain parallel visual axes to restore binocular single vision (BSV) when wearing the corrective prescription, thereby resulting in a fully accommodative esotropia, which may then be treated if indicated.

Surgery may be in the form of a unilateral medial rectus recession and lateral rectus resection, or bilateral medial recti recessions. DVD and associated muscle overactions may be corrected when performing the horizontal muscle surgery.

Refractive surgery laser-assisted in situ keratomileusis (LASIK) has been reported to correct refractive errors in older children and adults with a reduction in angle of deviation (Stidham *et al.* 2002). However, patients with pre-existing anisometropic amblyopia must be assessed carefully to ensure post-refractive surgery symptoms do not occur relating to poor fusion or a switch to fixation with the amblyopic eye (Calcutt & Manzouri 2006).

Constant esotropia without an accommodative element

Esotropia is present under all conditions and is unaffected by the state of accommodation. Presence of latent nystagmus, DVD, horizontal monocular asymmetry



Figure 6.2 Infantile esotropia.

of smooth pursuit and monocular asymmetry of optokinetic nystagmus (OKN) aid the diagnosis of early versus late onset strabismus (Schor *et al.* 1997).

Infantile esotropia (Fig. 6.2)

Infantile esotropia occurs in 0.1–1% of the population (Graham 1974, Nixon *et al.* 1985). It can be called the essential infantile esotropia syndrome because of its specific association with DVD and manifest latent nystagmus. Asymmetric motion visually evoked potential (VEP) and motion detection persists in infants who develop infantile esotropia, indicating a normal sensory capacity early on for these children that is later disrupted by abnormal binocular experience (Bosworth & Birch 2007).

Investigation

Case history

Infantile esotropia is associated with an early onset within the first 6 months of life (Friedrick & de Decker 1987, Nixon *et al.* 1985). Prematurity, low birth weight and low Apgar scores are significant risk factors for development of strabismus (Simonsz & Kolling 2011).

Refraction

There is often an associated degree of hypermetropia that requires long-term follow-up (Birch *et al.* 2010).

Abnormal head posture

There may be a face turn to adduct the fixing eye.

<i>Visual acuity</i>	There is a low incidence of amblyopia due to alternation of the deviation. Amblyopia is more common in infantile esotropia after treatment (Costenbader 1961, von Noorden 1988a, Shauly <i>et al.</i> 1994) but not before (Calcutt & Murray 1998, Friedman <i>et al.</i> 1980).
<i>Cover test</i>	A large-angle alternating esotropia is demonstrated and there may be associated DVD and latent nystagmus, which are usually detected after the age of 2 years (Lang 1968).
<i>Ocular motility</i>	Cross fixation is characteristic and limited abduction may be noted.
	Stimulation of the vestibulo-ocular reflex by swinging is useful in assessing the degree of abduction in a young infant. Cross-fixation occurs where the patient uses the right eye to fix in the left field of gaze and uses the left eye to fix in the right field of gaze.
<i>Optokinetic nystagmus</i>	This response is often asymmetrical with an absent or defective nasal to temporal response (Mein 1983, Demer & von Noorden 1988).
<i>Binocular function</i>	Patients are usually too young for assessment of retinal correspondence but in older patients this may be assessed by correcting the angle of deviation either in free space or using the synoptophore. Measurement of the deviation is most often assessed by corneal reflections in this younger age group. The measurement often exceeds 30 prism dioptres.
<i>Angle of deviation</i>	
<i>Associated syndromes</i>	Congenital strabismus syndrome (Lang 1968) includes esotropia, latent nystagmus and DVD.
<i>Differential diagnosis</i>	Ciancia syndrome includes infantile esotropia, latent nystagmus, abnormal head posture to adduct the eye and apparently limited abduction in either eye (Helveston 1993). Infantile esotropia must be differentiated from nystagmus block syndrome, accommodative esotropia, sixth nerve palsy, Duane's retraction syndrome, secondary esotropia, decompensated esophoria and late onset esotropia.

Nystagmus block esotropia

This is a condition in which manifest nystagmus is 'blocked' either totally or partially by placing one or both eyes in an adducted position. The presence of nystagmus in the primary position is often not detectable.

Investigation

<i>Case history</i>	An early onset within the first 6 months of life is typical. Esotropia develops as the patient constantly adducts the fixing eye to compensate for congenital nystagmus leading to constant esotropia. There is often an abnormal head posture to maintain one eye in adduction (Dell'Osso <i>et al.</i> 1983).
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<i>Visual acuity</i>	Amblyopia is common.
<i>Abnormal head posture</i>	Face turn to adduct fixating eye.
<i>Cover test</i>	A variable large-angle esotropia is present and is usually unilateral although both eyes often appear to be convergent.
<i>Ocular motility</i>	An increase in the amplitude of nystagmus on lateroversions is seen. On attempted abduction, the nystagmus often becomes apparent.
<i>Angle of deviation</i>	It is difficult to measure the angle with a prism bar as the eye continues to adduct on using base-out prisms. The position of corneal reflections is assessed using Hirschberg's method to estimate the angle of deviation.

Late onset (normosensorial) esotropia

Late onset esotropia occurs after a period of normal development of BSV.

Investigation

<i>Case history</i>	Onset is from the age of 2–4 years. The deviation may be intermittent for a short period of time but soon becomes constant and diplopia is appreciated by the older age group (Lang 1981, 1986). It may occur after temporary occlusion of one eye resulting in disruption of fusion or may be spontaneous and may be precipitated by illness. Uncorrected refractive error may be detected (Legmann Simon & Borchert 1997).
<i>Visual acuity</i>	There is a high incidence of amblyopia, if left untreated (Kothari 2007).
<i>Cover test</i>	A large angle of deviation is demonstrated.
<i>Binocular function</i>	Normal retinal correspondence and fusion are demonstrable on correction of the deviation.
<i>Prism adaptation</i>	The angle may not stabilise immediately with the prism but continue to increase at which stage, the target angle is stable with repeated measurements and associated with normal BSV at that angle (Savino <i>et al.</i> 2005).
<i>Differential diagnosis</i>	It is important to ensure that the deviation is not due to an acquired VI nerve palsy. The optic discs should be examined to exclude the presence of papilloedema.

Esotropia with myopia

This occurs in a small percentage of patients with non-accommodative esotropia. It has been proposed that binocular convergence develops so that the patient may use the far point, but is not relaxed on distance viewing resulting in esotropia. The deviation then becomes constant and progressively increases in angle. Esotropia

and hypotropia may also develop in high unilateral or bilateral myopia – this is discussed in Section ‘Hypotropia’.

Investigation

<i>Case history</i>	This esotropic deviation may present in childhood but is more common in adults and diplopia may be appreciated (Hugonnier & Clayette-Hugonnier 1969).
<i>Refraction</i>	A high degree of myopia is detected.
<i>Visual acuity</i>	The level of acuity is often affected by degenerative myopic fundus changes.
<i>Cover test</i>	The deviation progressively increases on dissociation.
<i>Ocular motility</i>	An associated limitation of abduction may be noted.
<i>Binocular function</i>	Normal retinal correspondence and fusion should be demonstrable on correction of the deviation but is often difficult to assess due to the reduced levels of visual acuity and the marked angle of deviation.
<i>Differential diagnosis</i>	It is necessary to ensure that the deviation is not due to thyroid eye disease as both have prominent eyes and may show limited abduction.

Management of constant non-accommodative esotropia

Correct the refractive error and treat amblyopia if present. Surgery may be for a functional result with improved ocular alignment, widened peripheral field of view and psychosocial impact or a functional result with attainment of binocular vision.

Congenital/Infantile: Aim for parallel visual axes. It is often advocated that if surgery is performed before 2 years of age, there is a stronger potential for achieving binocular vision (normal retinal correspondence or, more commonly, abnormal retinal correspondence) (Ing 1983, Parks 1984). Early surgery is also associated with lack of development of DVD and latent nystagmus plus development of symmetrical OKN (Wong *et al.* 2008). Decision to wait until the child is older to perform surgery relates to the ability to achieve accurate measurements, detailed investigation, full development of the eye and visual pathway and allows combined surgery where DVD and/or inferior oblique overaction occurs. The ELISSS (early versus late infantile strabismus surgery study) reported better sensory outcomes with early surgery but less operations required for the late surgery group (Simonsz *et al.* 2005). Reoperation rates are reported as 60–80% for children first operated around age of 1 year and 25% for those operated around age 4 years (Simonsz & Kolling 2011).

The procedures may involve bilateral medial rectus recessions, or bilateral medial rectus recessions combined with unilateral lateral rectus resection if there is a large angle deviation (Minkoff & Donohue 2005). Hang-back recession is reported to be as effective as conventional medial rectus recession (Spierer & Spierer 2010). Surgery may also include use of faden sutures (Rizk 1999). However, treatment is usually with bilateral medial recti recessions (Helveston *et al.* 1983, Ing *et al.* 1966,

Kushner & Morton 1984, von Noorden *et al.* 1972, Prieto-Diaz 1980, 1985, Weakley 1991) and this allows later surgery on the lateral rectus muscles and oblique muscles as required (Keskinbora & Pulur 2004). A randomised controlled trial has shown no difference between results of bilateral medial rectus recessions versus unilateral lateral rectus resection with medial rectus recession (Polling *et al.* 2009). Where there is coexistent A or V pattern, vertical transposition of the medial rectus muscle may be undertaken (Dickmann *et al.* 2011).

Botulinum toxin has been used as a sole therapeutic intervention with some success in achieving long-term alignment of the visual axes (Campos *et al.* 2000, McNeer *et al.* 2000, McNeer *et al.* 2003, McNeer & Tucker 2010). One trial reports equivalent success of botulinum toxin versus surgery in retreatment for infantile esotropia (Tejedor & Rodriguez 1999).

Nystagmus block syndrome: Surgery is unpredictable. A similar procedure to that in infantile esotropia is employed (von Noorden & Wong 1986).

Normo-sensorial: Aim for parallel visual axes or overcorrect slightly as a functional result is indicated. Botulinum toxin can provide equally good outcomes versus surgery for retreatment of children with acquired esotropia (Tejedor & Rodriguez 1998). The longer the duration of misalignment before surgery, the worse the effect on binocular outcomes (Kaseem & Elhilali 2006).

Esotropia with myopia: Disappointing results are common as the deviation tends to recur.

In intermittent esotropia, the esotropia is only present under certain conditions.

Accommodative esotropia

The convergent deviation is affected by the state of accommodation and this is the primary factor in the aetiology of the squint.

Fully accommodative esotropia (Fig. 6.3)

BSV is present for all distances when hypermetropia is corrected.

Investigation

Case history An intermittent onset is usual between the ages of 2–5 years. It may be precipitated by illness and there is often a positive family history for squint and/or hypermetropia.

Refraction There is a moderate degree of hypermetropia and may also have a degree of astigmatism and/or anisometropia. Ensure that full prescription is given as deviation may remain intermittently manifest with glasses otherwise.

Visual acuity Amblyopia is uncommon but may be present if the refractive correction is not worn until after a unilateral deviation has become well established.

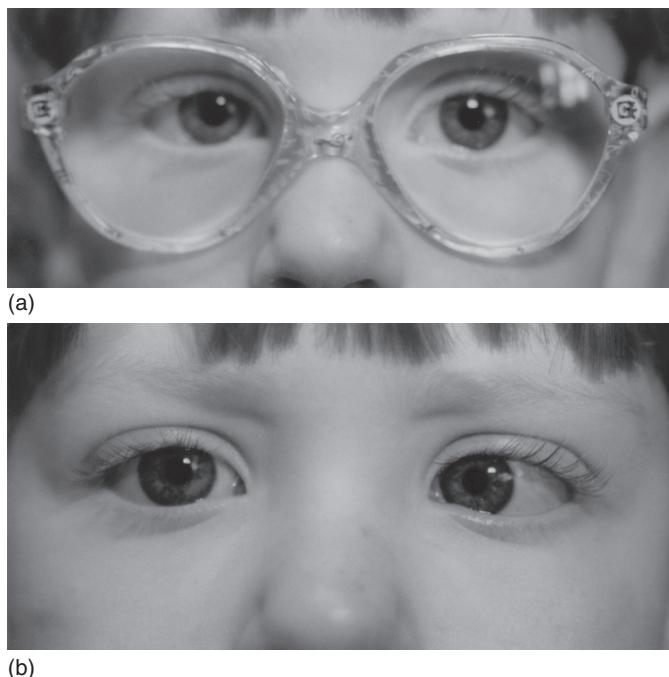


Figure 6.3 Intermittent fully accommodative esotropia.

<i>Cover test</i>	A manifest deviation is seen without the spectacle correction but is controlled to a latent deviation with the full hypermetropic correction.
<i>Binocular function</i>	BSV is demonstrated for near and distance fixation when fully corrected. Usually, normal retinal correspondence is present unless the deviation is controlled to a microtropia where there is anisohypermetropia and a central suppression scotoma. Without glasses, there is suppression with the manifest esotropia. Older children may be aware of the esotropia without glasses and appreciate 'misty and clear'. Binocular visual acuity shows good control at near and distance with glasses but reduced without glasses. Corrected binocular acuity assessment may show increase in the esotropia if the patient has greater difficulty fusing small targets than large targets whilst having to accommodate extra. This is expected to be normal.
<i>AC/A ratio</i>	A slight convergent angle is measured with glasses increasing without glasses and on accommodation.
<i>Angle of deviation</i>	This demonstrates a central suppression scotoma when there is a microtropia with the spectacles. Where there is a latent deviation with the spectacles, there is bifoveal fixation.
<i>Four dioptre prism test</i>	

Management

Give the full refractive correction after cycloplegic refraction. Hypermetropia may be seen to have increased on the second refraction and the deviation often will have become more noticeable when the spectacles are removed. Children who are given their full correction show smaller and more controlled deviations than those undercorrected (MacEwen *et al.* 2009). It is also likely that many patients will require glasses into adulthood (Mohney *et al.* 2011).

Treat amblyopia if present. Orthoptic exercises may be indicated with older children to control the manifest deviation. It is important to take into account the strength of the correction.

Treatment for patients with a spectacle correction of < +3.0 DS and +1.0 cyl. Children with lower corrections show good outcomes from being weaned out of glasses (Lambert *et al.* 2003). Aim to have good BSV and binocular visual acuity, and normal accommodation and convergence without the spectacle correction. Anti-suppression exercises may be indicated initially to allow appreciation of pathological diplopia without glasses. Concave lenses and bar reading may be used to improve negative relative convergence with stereograms (distance fixation). Fusion and synoptophore exercises may be used in addition in the clinical setting. Teach the patient to relax accommodation to gain one blurred image without the spectacles (misty). Then gradually encourage accommodation while maintaining the one image (clear). Spectacles may be left off under supervision at a later stage but prior to that, the strength of the correction may be gradually reduced.

Treatment for patients with a spectacle correction of > +3.0 DS and +1.0 cyl. The patient will always need some hypermetropic correction, particularly for close work.

Exercises may be given to achieve ‘social’ control of the deviation for sports and photographs but not for close work, etc. The aim is to obtain one misty, blurred image with relaxed accommodation. Two clear images will be seen on accommodation. Exercises may also be given to improve control of the deviation in order to reduce the strength of correction in spectacles for cosmetic reasons.

Surgery is not usually indicated in this type of deviation. However, it may be considered if there is a persistent large angle deviation despite wearing the full correction with a risk of decompensation. Approximately 10% are reported as eventually requiring strabismus surgery and these cases are associated with earlier onset and diagnosis of the strabismus (Mohney *et al.* 2011).

Refractive surgery (e.g. LASIK) has been reported to correct refractive errors in older children and adults with a reduction in angle of deviation (Stidham *et al.* 2002). It is recommended that refractive surgery is not performed in younger children as they have greater amounts of latent hypermetropia and thus, may become strabismic again as refractive error changes in older life (Kirwan *et al.* 2010).

Convergence excess esotropia (Fig. 6.4)

There is BSV for distant fixation, but esotropia on accommodation for near fixation.



Figure 6.4 Intermittent convergence excess esotropia.

Investigation

Case history

Onset is usually between the ages of 2 and 5 years and is noticed when the patient is looking at close objects. However, if this type of squint is associated with hypermetropia that has not been corrected, the deviation will have been noticed for both near and distance. There is often a positive family history.

Refraction

The deviation may be associated with hypermetropia and occasionally a myopic refractive error is detected (Calcutt 1983, 1989).

Cover test

A manifest deviation is present for near fixation on accommodation but control may be demonstrated with a light target as less or no accommodation is exerted. A latent deviation is present for distance fixation.

Convergence

May be binocular to a light target but not to an accommodative target. Overconvergence may occur.

Accommodation

Where a reduced amplitude of accommodation is found, a diagnosis of hypoaccommodative convergence excess esotropia must be considered.

<i>Binocular function</i>	BSV is present on distance fixation and can be assessed using the synoptophore or free-space distance tests. A spotlight target and non-dissociative tests may demonstrate binocularity for near fixation. Normal retinal correspondence is usually present unless the deviation is controlled to a microtropia. Binocular visual acuity is normal for distance and reduced or absent for near fixation.
<i>Angle of deviation</i>	A slight convergent deviation is measurable at distance fixation with a larger manifest angle at near fixation.
<i>Prism adaptation</i> +3.0 DS	This will elicit the maximum angle of deviation for near fixation to aid with surgical planning (Leuder & Norman 2006). The deviation is reduced by eliminating the need to accommodate for near fixation – esophoria is seen. BSV may be proved at this stage with Bagolini glasses and stereotests.
<i>AC/A ratio</i>	A high ratio at greater than 5:1 is expected.
<i>Monocular occlusion</i>	A period of monocular occlusion of at least 30 minutes may alter the angle of deviation and AC/A ratio by eliminating functional tenacity (Garretty 2010).
<i>Differential diagnosis</i>	It is necessary to ensure the deviation is not a constant accommodative type esotropia, an intermittent near type esotropia, intermittent fully accommodative esotropia with uncorrected or undercorrected hypermetropia or due to sixth nerve palsy or superior oblique underaction with a V eso pattern.
	Hypo-accommodative esotropia is greater at near than distance fixation, is unrelated to an uncorrected hypermetropic refractive error and is caused by excessive convergence from an increased accommodative effort to overcome a primary or secondary weakness of accommodation. It is due to a remote near point of accommodation (Calcutt & Kinnear 1997, James 1997).

Management

Correct any refractive error and treat amblyopia if present.

Surgery is required in many cases, particularly if there is a high AC/A ratio (Leuder & Normal 2006).

The aim is to reduce the near angle of deviation so that BSV is achieved, without overcorrecting the smaller distance angle, thus affecting binocularity for distance. Surgery reduces the near angle and collapses the near-distance disparity (Arnoldi & Shainberg 2005). The procedure usually involves bilateral medial rectus recessions and/or medial rectus posterior fixation sutures (fadens) depending on the size of the deviation (Leitch *et al.* 1990) or slanting suturing of medial rectus recessions (Gharabaghi & Zanjani 2006). Combined research/recession of the medial rectus muscle specifically targets the near-distance disparity (Ramasamy *et al.* 2007).

Orthoptic exercises may be used with older cooperative children. The deviation should be less than 20 prism dioptres in angle with an AC/A ratio of no more than

8:1 and some appreciation of control for near fixation already. Anti-suppression exercises may be necessary to allow appreciation of pathological diplopia. Accommodation and negative relative convergence are further improved to achieve good binocularity for near fixation. Accommodation is relaxed to obtain one misty image, and then accommodation gradually encouraged while maintaining one clear image.

Bifocals may be used with older children (usually over 5 years of age). Straight-topped executive bifocals are used ensuring that the near segment bisects the pupil (Arnoldi 1999, Vivian *et al.* 2002). The weakest addition (up to +3.0) is given to allow comfortable BSV and good binocular visual acuity. The aim is to restore to single vision lenses as soon as possible by reducing the bifocal segment strength over the follow up period (Burian 1956). Long-term wear of bifocals can induce hypoaccommodation (Fresina *et al.* 2010).

Miotics act directly on the ciliary muscles by causing a spasm of the ciliary body, which induces increased lens convexity (Chatzistefanou & Mills 2000). Miosis gives an increased depth of focus so that less accommodation is required. Phospholine-iodide 0.06/0.125% has been used in past years. This is no longer widely available in the UK market but may be available on a named patient basis from pharmacy departments for use under expert supervision. Generalised parasympathomimetic side effects such as sweating, bradycardia and intestinal colic may follow systemic absorption of these eye drops. Other effects may include hypersalivation and bronchospasm (Abraham 1961, Decker 1972). Iris cysts may be a local complication (Hiatt 1983). Pilocarpine 4.0% may also be used but is not found to be as satisfactory as it has to be instilled on a more frequent basis. If after 1 month there is no improvement of control, the miotics are discontinued. If improvement has occurred, continue for 2–3 months and then gradually wean off over several weeks.

Contact lenses: The equivalent strength contact lenses to the spectacle correction are given initially. An additional 1 D may be given if the esotropia persists. These are useful with patients with high AC/A ratios and those with associated myopia (Calcutt 1983, 1989).

Relating to fixation distance

Near esotropia

BSV is present for distance fixation and esotropia for near fixation even when the accommodative effort is relieved.

Investigation

- | | |
|---------------------|--|
| <i>Case history</i> | An early onset is usual. Diplopia is noted if occurring in older patients. Patients may complain of asthenopic symptoms if trying to maintain control. |
| <i>Refraction</i> | There may be associated hypermetropia. |

<i>Visual acuity</i>	Patients tend not to be amblyopic as there is no manifest deviation at distance.
<i>Cover test</i>	A manifest deviation for near fixation is present both with and without accommodative stimulus. A latent deviation is present for distance fixation.
<i>Ocular motility</i>	Bilateral superior oblique weaknesses may be noted.
<i>Convergence</i>	Absent or reduced convergence is recorded unless there is intersection of the visual axes followed by symmetrical convergence.
<i>Binocular function</i>	Normal retinal correspondence is demonstrable but binocular functions are weak. Binocular visual acuity is normal for distance and reduced or absent for near fixation.
<i>Angle of deviation</i>	Slight convergent deviation at distance and larger manifest angle at near fixation.
<i>AC/A ratio</i>	Normal. The deviation is unaffected by +3.0 DS lenses and there is a normal accommodative near point (von Noorden & Avilla 1986).
<i>Differential diagnosis</i>	A bilateral fourth nerve weakness should be excluded by careful examination of the ocular movements.

Management

Correct any refractive error and treat amblyopia if present. Orthoptic exercises are contraindicated as near esotropia is not an accommodative deviation, and lenses and miotics are of limited help. Surgery, if indicated, usually involves bilateral medial recti recessions and/or fadens depending on the angle of deviation (von Noorden & Avilla 1986).

Distance esotropia

Esotropia is present on distant fixation with BSV present on near fixation.

Investigation

<i>Case history</i>	Diplopia is noted by older patients.
<i>Visual acuity</i>	Amblyopia is uncommon.
<i>Cover test</i>	A manifest deviation is present for distance and a latent deviation for near fixation.
<i>Ocular motility</i>	It is important to rule out lateral rectus weakness.
<i>Binocular function</i>	Suppression responses are recorded at distance fixation but BSV when the angle is corrected on the synoptophore. Good fusion and stereopsis responses are recorded at near fixation using free space tests. Binocular visual acuity is normal for near and reduced for distance fixation.

<i>Angle of deviation</i>	Slight convergent deviation at near and larger manifest angle at distance fixation.
<i>Differential diagnosis</i>	Ensure the deviation is not due to mild VI nerve paresis, thyroid eye disease with slightly limited abduction, convergence and accommodative spasm or divergence palsy.

Management

Treat amblyopia if present.

Prisms may be used with adult patients where a small angle of deviation, less than 10 prism dioptres, is present for distance and the patient wears spectacles (Mittelman 2006).

Surgery, if indicated, usually involves bilateral lateral recti resections of equal but small amount. Bilateral medial rectus recessions have also been utilised for this type of deviation (Bothun & Archer 2005).

Cyclic esotropia

This deviation relates to time with the deviation occurring at regular intervals.

Investigation

<i>Case history</i>	There is usually a late onset from 4 to 5 years. The most common pattern is an alternate day deviation. After a variable period of time, the alternate day pattern may alter and the deviation eventually becomes constant (Rowe 1995).
<i>Visual acuity</i>	Amblyopia occurs in approximately 20% (Cahill <i>et al.</i> 1999).
<i>Cover test</i>	On the squinting day, a marked deviation for near and distance fixation is present. On the straight day, a latent deviation for near and distance fixation is present, which is usually esophoria (Cahill <i>et al.</i> 1999).
<i>Binocular function</i>	Suppression is present on the squinting day with normal BSV present on the straight day. On a straight day, there is normal retinal correspondence with good fusion and stereopsis. On a squinting day, there is suppression but normal retinal correspondence is evident on correcting the angle of deviation in free space or using the synoptophore.

Management

Surgery is usually indicated for the manifest deviation. This may be performed once the deviation has become constant but may also be performed before the deviation has altered its cyclical pattern. As the deviation is often unilateral, a medial rectus recession and lateral rectus resection procedure on the deviating eye is usual. There is a good prognosis for restoration of BSV (Parlato *et al.* 1983).

Non-specific esotropia

An intermittent esotropia not conforming to any pattern.

Investigation

<i>Case history</i>	Older patients may be symptomatic and complain of headaches and intermittent diplopia.
<i>Cover test</i>	The deviation is intermittently manifest at any fixation distance and there is no significant change in the angle of deviation for near or distance fixation or with accommodation exerted.
<i>Binocular function</i>	Normal retinal correspondence is present but weak.
<i>Differential diagnosis</i>	Decompensating esophoria.

Secondary esotropia

Esotropia that follows loss or impairment of vision.

Investigation

<i>Fundus and media</i>	Retinal changes related to the cause of impaired vision may be examination evident.
<i>Case history</i>	Children who present with secondary esotropia indicate an onset of the visual loss or impairment from the age of 6 months to 7 years (Havertape <i>et al.</i> 2001). Adults who present with secondary esotropia indicate the presence of a pre-existing esophoria. Severe visual loss is present, which may be unilateral or bilateral. Unilateral esotropia with poor fixation is usual.
<i>Visual acuity</i>	Limitations of motility may be noted if there is associated damage to the globe and extraocular muscles.
<i>Cover test</i>	This is usually measured using corneal reflections, as visual acuity is too poor to fixate.
<i>Ocular motility</i>	Poor fixation is noted.
<i>Angle of deviation</i>	This is absent or reduced, dependent on the extent of visual loss.
<i>Visuscope</i>	
<i>Binocular function</i>	
<i>Suppression</i>	Post-operative diplopia risk must be evaluated in patients over 7 years of age.

Management

Treat the cause of visual loss if possible. Surgery is usually for corrective reasons. The aim is to leave the deviation slightly undercorrected as it should further decrease with time and this may guard against a later onset of consecutive exotropia.

Botulinum toxin is a well-tolerated treatment option and the angle of deviation often reduces with repeated injections (Dawson *et al.* 2005).

Consecutive esotropia

Esotropia in a patient who has previously had exotropia or exophoria.

Investigation

<i>Refraction</i>	There may be under or uncorrected hypermetropia, which may be beneficial to the esotropia when corrected fully.
<i>Case history</i>	There is often a history of previous surgery that may have been deliberate to prevent redivergence in certain cases. The patient may appreciate diplopia immediately post-operatively, which usually resolves. Spontaneous onset of consecutive esotropia is rare (Fitter & Jampolsky 1964)
<i>Visual acuity</i>	Patients may be amblyopic if unilateral esotropia has developed or if amblyopic from previous exotropia.
<i>Ocular motility</i>	There may be a limitation of ocular movements, particularly in cases of unplanned surgical overcorrection.
<i>Binocular function</i>	Where binocular function was previously demonstrated pre-operatively, this should remain immediately post-operative. However, suppression is more often detected.

Management

When aiming for a functional response, if the deviation has not resolved within 2 weeks of surgery, post-operative treatment should be undertaken. Amblyopia may develop, which requires treatment.

Convex lenses: Small angle deviations may respond to use of convex lenses, particularly if there is a high AC/A ratio.

Orthoptic exercises may be given to improve negative relative convergence.

Prisms may be used to achieve BSV and are gradually reduced at a later stage while maintaining binocularity.

Miotics may be used in cases of accommodative esotropia to reduce accommodation by increasing the depth of focus.

When a corrective result is required, *surgery* may be indicated depending on the angle of deviation and the appearance of the deviation.

When a cosmetic result is required, *surgery* may be indicated depending on the angle of deviation and the cosmetic appearance.

Exotropia

A condition in which one or other eye deviates outwards.

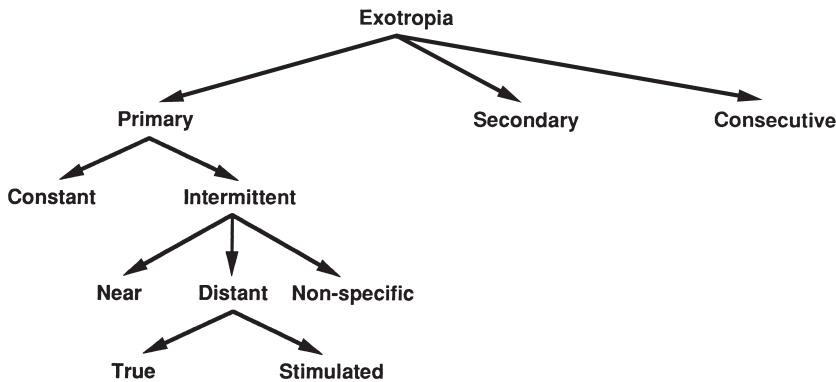


Figure 6.5 Classification of exotropia.

Classification

See Fig. 6.5.

Aetiology

Primary

Constant. Anatomical (wide IPD, exophthalmos, orbital asymmetry, muscle anomalies, craniofacial abnormalities), hereditary, myopia

Intermittent.

- *Near:* Acquired myopia, presbyopia
- *Distant:* High AC/A ratio, anatomical
- *Non-specific:* Any of the above, weak fusion

Secondary

Loss or impairment of vision (birth or >8 years)

Consecutive

Surgical overcorrection, passage of time

Primary exotropia

The divergent deviation constitutes the initial defect.

Constant exotropia (Fig. 6.6)

The exotropia is present under all conditions.

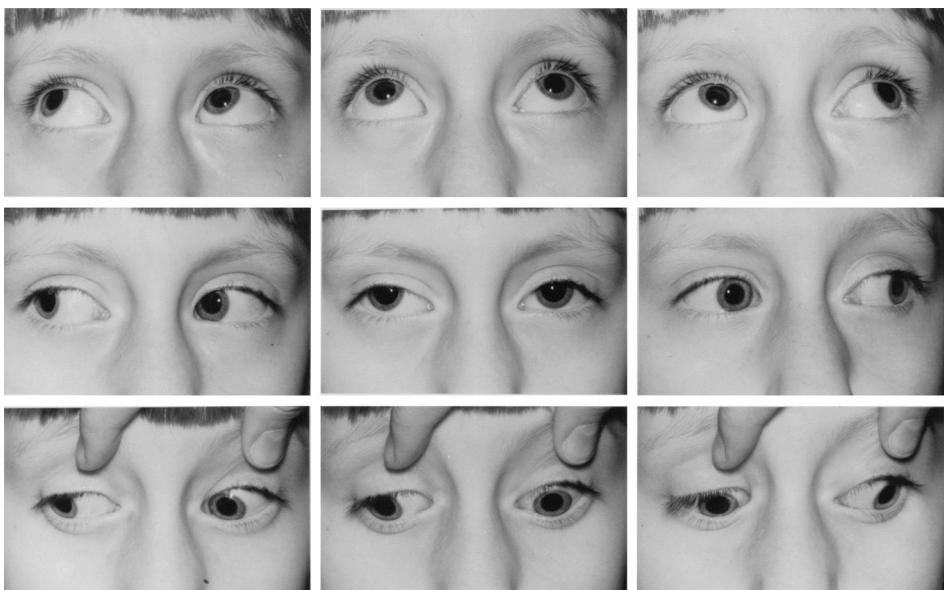


Figure 6.6 Constant exotropia.

Investigation

Case history

The deviation may have been intermittent first and then became constant; older children and adults may present like this. With early onset exotropia in children, there may be a history of closing one eye in sunlight. The patient may be aware of panoramic vision.

General health; exotropia may be present in young children with slow mental and physical development and may decrease as general development improves.

This is often equal if the deviation is alternating.

There is a moderate angle exotropia at all distances, which often alternates. DVD may be noted if infantile in onset.

A V exo pattern may be documented.

Convergence to 6 cm with the same angle of deviation indicates ARC. Convergence with reducing angle of deviation to intersection of the visual axes followed by symmetrical convergence of either eye indicates normal retinal correspondence. Further divergence following intersection of the visual axes indicates absence of retinal correspondence. Retinal correspondence cannot be estimated where there is convergence to 6 cm with reducing angle of deviation as intersection of the visual axes does not occur.

Visual acuity

Cover test

Ocular motility

Convergence

Accommodation

This may be reduced, particularly if there is a late onset exotropia. The decreased amplitude of accommodation may contribute to the decompensation of the deviation.

Binocular function

Free space tests may be used to assess the presence of ARC. A binocular response to Bagolini glasses or Worth's 4 lights is indicative of normal or ARC but ARC will be seen in small angle deviations rather than large angles. Presence of ARC is unusual in exotropia but may develop in early onset exotropia, e.g. infantile. Normal retinal correspondence is noted in later onset exotropia. Free space fusion and stereopsis may be further tested to examine the quality of ARC if present. Suppression responses on free space tests do not give an indication of retinal correspondence. Suppression is often quite dense.

Retinal correspondence may be evaluated in full using the synoptophore. Normal retinal correspondence is present where superimposition is achieved at the full-corrected angle of deviation. ARC is present where superimposition is achieved at the angle of deviation. Quality of retinal correspondence, whether normal or abnormal, may be determined by assessing the fusion and stereopsis responses at these angles (Moore & Cohen 1985, Burian 1945).

Where suppression responses are obtained, it is not possible to determine retinal correspondence. Therefore, further assessments may be undertaken including afterimages and using vertical prisms to move the image out of the suppression area and align the vertically displaced images using horizontal prisms. Where there is suppression and the patient is aged 7 years or greater, the depth and area of suppression should be measured in order to assess the risk of post-operative diplopia.

Angle of deviation

This should be measured for near and distance fixation.

Management

Classically, correcting hypermetropia will improve an eso-deviation and worsen an exo-deviation (Swan 1983). However, if there is significant blurring of vision, correcting hypermetropia will improve fusional convergence and lessen the amount of exo-deviation (Iacobucci *et al.* 1993). Treat amblyopia if present.

Corrective result. Use of botulinum toxin A may be considered if there is a possibility of post-operative diplopia. This enables the deviating eye to take up a straighter position by temporally paralysing the medial rectus muscle and allowing a full assessment to be made of potential binocular function or the post-operative

risk of diplopia. Some patients prefer to undertake repeated botulinum toxin A injections in preference to undergoing corrective surgery.

Surgery, if indicated, is usually a unilateral recession and resection procedure, using adjustable sutures as required with older patients (Khazaeni & Volpe 2006). The aim is for parallel visual axes or a slight overcorrection. A positive correlation has been reported for preoperative distance angle of deviation and response to surgery with better response to unilateral recession/resection surgery (Faridi *et al.* 2007). For small angles, unilateral medial rectus resection is effective (De Decker & Baenge 1988).

Functional result. Use of adjustable suture surgery is again preferable with the older patient with the aim for parallel visual axes or a slight undercorrection. For small angles of deviation, a prism may be used to achieve comfortable BSV.

In intermittent exotropia, the exotropia is only present under certain conditions.

Near exotropia

Exotropia is present on near fixation and BSV on distant fixation.

Investigation

<i>Refraction</i>	Patients may be myopic. If hypermetropia is present, order only the minimum prescription to maintain adequate visual acuity.
<i>Case history</i>	A later onset is common, usually with onset of myopia or presbyopia. Diplopia and asthenopic symptoms may be appreciated.
<i>Visual acuity</i>	Corrected acuity is usually equal.
<i>Cover test</i>	A manifest deviation is present for near fixation and a latent deviation for distance. Patients may have an exophoria to accommodative target at near fixation but exotropia to a light source.
<i>Convergence</i>	The level of convergence is poor or absent as a result of the divergent angle at near fixation.
<i>Binocular function</i>	This can be assessed using the synoptophore or free space tests at distance. Normal retinal correspondence with demonstrable fusion involving a poor positive range is noted. For near fixation, there may be suppression or crossed pathological diplopia that is joined with the appropriate prismatic correction.
<i>Angle of deviation</i>	Slight divergent angle for distance increasing for near fixation.
<i>Differential diagnosis</i>	It is necessary to ensure that the deviation is not due to convergence palsy or convergence insufficiency.



Figure 6.7 Intermittent distance exotropia.

Management

Correct the refractive error as appropriate.

Orthoptic exercises may be used in cases where the angle of deviation measures less than 20 prism dioptres. Anti-suppression exercises may be necessary to enable appreciation of pathological diplopia. The positive fusional range is improved with prism and synoptophore exercises and the positive relative convergence is improved with stereograms (near fixation). Binocular near point of convergence is improved with convergence exercises.

Base-in prisms and/or concave lenses may be used as a support to orthoptic treatment. The minimum strength of prism is given to control the deviation and the aim is to reduce the strength as soon as possible (Hardesty 1972). Concave lenses are seldom useful for long-term wear as they may blur visual acuity.

Surgery if indicated may involve bilateral medial rectus resections. It is advisable to do a small amount bilaterally than a large unilateral resection. Alternatively, a small lateral rectus recession with larger medial rectus resection may be undertaken (Choi *et al.* 2007).

Diagnostic corrective prisms may be used to assess the effect on distance fixation pre-operatively.

Orthoptic exercises to improve positive relative convergence may be given post-operatively to consolidate the result.

Distance exotropia (Fig. 6.7)

Exotropia is present on distant fixation and BSV on near fixation. The angle of deviation for near fixation may increase on prolonged disruption of fusion or elimination of accommodation. Cases that show such increase for near fixation are simulated distance types. The natural history of distance exotropia shows that it is generally a progressive strabismus type.

Investigation

<i>Refraction</i>	May be myopic. If hypermetropic, the minimum correction should be ordered.
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Case history

The age of onset of many exo-deviations (constant and intermittent) is early and usually before the age of 2 years (Biglan *et al.* 1996, Hall 1961). Small angle deviations may not present until later because of the intermittent nature of the deviation. The deviation is noticed when daydreaming or tired. The patient is usually asymptomatic but may be aware of the divergence from the feel of the eye or increase in the visual field. The deviation is often worse during the summertime and photophobia appreciated. The eyes may also tend to diverge in bright sunlight and the patient may close one eye in such circumstances (Holt 1951, Berg 1982). This is due to the increased depth of focus, which is known to occur with miosis pupils. As a result, less accommodation and therefore less accommodative convergence is required, resulting in decompensation of the exo-deviation. The deviation tends to occur more frequently with poor health. The patient may complain of blurred vision if using accommodation to control the deviation in the distance and may be aware of micropsia if using excessive accommodation to control the deviation. Control may vary throughout the day being better in the morning; therefore, it is useful to vary appointment times to assess overall ability to control the deviation.

This is usually equal.

Visual acuity

There is a latent exo-deviation on near fixation with a manifest deviation for distance and far distance. The deviation may appear controlled initially for distance but compensates quickly on dissociation.

Cover test

Motility is often full, but patients may have a V exo pattern with slight bilateral superior recti underactions. Lateral incomitance may be measured whereby the divergent angle of deviation reduces on lateral gaze (Moore 1969, Caldeira 1992). Compare the alternate cover test in primary position with right and left gaze positions. A reduction of more than 20% of the deviation is regarded as significant (von Noorden 1995). With lateral gaze incomitance, a decrease in the amount of lateral rectus recession is required (Meyer 1981, Moore 1969).

Ocular Motility

This is usually good, but if reduced this may be an early sign of decompensation.

Convergence

Monocular and binocular near points may be measured.

Accommodation

<i>Binocular Function</i>	Normal retinal correspondence with fusion and stereopsis is present at near fixation. Note the base-out range, as fusional convergence is important in maintaining control for near. Suppression or pathological crossed diplopia is present for distance that is joined with the appropriate prismatic correction. Suppression may be a hemiretinal suppression (Jampolsky 1955) or a less extensive suppression scotoma, extending over the nasal and temporal retina similar to suppression in esotropia (Pratt-Johnson & Tillson 1984). Anti-diplopia mechanisms appear to be triggered by images falling on temporal retina versus nasal retina (Serrano-Pedraza <i>et al.</i> 2011).
<i>Angle of deviation</i>	Binocular visual acuity is good for near acuity and reduced for distance. The near binocular visual acuity may be reduced in cases of simulated distance exotropia due to accommodative convergence control. The eyes may tend to diverge when accommodation is relaxed in order to see the smaller letters during the test.
<i>Control score</i>	A slight divergent angle is measured for near fixation, which may increase on dissociation. A larger angle is measured for distance and far distance fixations. The deviation should be measured for near, distance, far distance, on lateral gaze and in vertical gaze positions.
<i>AC/A ratio and +3.0 DS</i>	A calculation of control can be made for intermittent exotropia based on quantitative measures combined with qualitative assessment of how much the eye deviates. The Newcastle control score (Buck <i>et al.</i> 2007, Buck <i>et al.</i> 2008, Clarke 2007, Haggerty <i>et al.</i> 2004) combines values from home plus near and distance assessments to give a score ranging from 0 (full control) to 9 (no control). Home represents the percentage time the deviation is noted at home for distance and near fixation. The clinic measurements include rate of recovery of cover test at near and distance (immediate recovery, after blink or refixation, remains manifest after dissociation or prolonged fixation, manifest spontaneously).
	Variability in control may also occur throughout the day and control is not always worse towards the end of the day (Hatt <i>et al.</i> 2008).
	If simulated due to accommodative convergence, the angle of deviation will increase to equal or more than the distance measurement when measured for near fixation with +3.0 DS lenses in place. The AC/A ratio will be high if the deviation is controlled by accommodative convergence, indicating a simulated distance exotropia. Check to ensure that the target can be seen clearly with the lenses in place.

An increase in angle greater than 10 prism dioptres will differentiate between true and simulated distance exotropia in cases associated with accommodation.

Diagnostic occlusion

If simulated due to strong fusional convergence hold, the angle of deviation will increase for near fixation after a period of monocular occlusion (at least $\frac{1}{2}$ hour). The deviation should increase to equal or near equal to the distance measurement. It is important to ensure that the patient never has a chance to regain BSV when removing the occlusion to perform the prism cover test (Walklate 1998). The deviation is unlikely to increase following diagnostic occlusion if the control of the deviation is already poor (Arnoldi & Reynolds 2008).

Management

Correct the refractive error. Correcting a myopic error is particularly useful in aiding control of the exo-deviation. A hypermetropic error is undercorrected where possible but ensuring good visual acuity is maintained. Most children with well-controlled exotropia receive no treatment within the first 12 months following presentation (Buck *et al.* 2009).

Orthoptic exercises may be useful in small angle deviations less than 15 prism dioptres. Anti-suppression exercises may be required initially to demonstrate pathological diplopia (Luke 1970). It is necessary to improve binocular convergence, fusional amplitudes and positive relative convergence. Orthoptic treatment is contraindicated in cases where there is a possibility of surgery at a later stage, as there would be a risk of overconvergence post-operatively.

Concave lenses, up to -4.0 DS, in addition to the underlying refractive correction may be used in cases with high AC/A ratio to stimulate accommodative/convergence and lenses are often combined with orthoptic exercises. The protocol is to provide the minimum minus lenses required to achieve control of the manifest deviation. This is then reduced by -0.50 DS steps every 6 months ensuring that binocular control is maintained. Fifty percent report a good outcome with this treatment (Caltrider & Jampolsky 1983, Kushner 1999, Rowe *et al.* 2009).

Base-in prisms may be used pre-operatively to obtain control and are often combined with orthoptic exercises. The aim is to reduce the strength of prism as soon as possible.

Alternating occlusion: Occlusion of either eye (2:1 or 5:2 dominant to non-dominant eye ratio) alternately for 6–12 weeks may improve sensory function and motor control. Alternate occlusion alters sensory input to the visual system, which reduces the suppression and improves fusional amplitudes. There is subsequent improvement in motor control with increased control of the deviation. Risks may include no improvement in suppression and loss of fusional amplitude, increase in angle of deviation and reduction in motor control (Flynn *et al.* 1975, Vishnoi *et al.* 1987).

Tinted spectacles: These are useful if bright light constitutes a major dissociative factor and may enable the patient to maintain control of the deviation in bright light (Eustace *et al.* 1973).

Surgery is required for large angle deviations and when decompensation of the deviation occurs for near fixation.

Indications for delaying surgery (Abroms *et al.* 2001, Asjes-Tyndeman *et al.* 2006) include the following:

- Observe progress of the deviation, particularly variable control of a distance deviation.
- Allow for full facial development.
- More accurate measurements may be obtained when older.
- Post-operative treatment may be carried out.

Reasons for early surgery (Chia *et al.* 2005) include the following:

- Prevents sensory changes.
- Minimises the development of a constant manifest deviation and deterioration of BSV.
- The cosmetic appearance may be poor.

Surgery should not be delayed if the deviation begins to decompensate for near and there is development of secondary convergence insufficiency (Pratt-Johnson *et al.* 1977). The aim of surgery is to achieve parallel visual axes or a slight overcorrection. It is important to take into account the presence of lateral incomitance as this may lead to overcorrection post-operatively, should lateral rectus restrictions occur. Surgery usually involves bilateral lateral rectus recessions for true distance exotropia and unilateral lateral rectus recession/medial rectus resection for simulated distance exotropia (Burian & Spivey 1965, Kushner 1998, von Noorden 1969). Bilateral lateral recti recessions may also be considered whatever the type of distance exotropia (Kushner 1998). Modifications to surgery can include allowance for lateral gaze incomitance and presence of V pattern exotropia.

Randomised and non-randomised trials show a higher success rate with recess-resection procedures (Chia *et al.* 2006, Jeong *et al.* 2006, Kushner 1998). A higher success rate is reported when the larger angle (after dissociation and on far distance measurement) is operated (Kushner 1998). Overcorrection is often planned because of the known risk of exo drift post-operatively with an aim for 10 PD eso angle but less than 20 PD (Kim *et al.* 2005, Koo *et al.* 2006).

Factors associated with recurrence of exo-deviation include amblyopia or difference in visual acuity (Koklanis & Georgievski 2008).

Botulinum toxin may be used as an alternative to surgery for post-operative overcorrections.

Cyclic alternate day exotropia

This deviation relates to time with the deviation occurring at regular intervals.

Investigation

<i>Case history</i>	There is usually a late onset from 4–5 years. The most common pattern is an alternate day deviation. There may be a prior history of intermittent exotropia (Dawson <i>et al.</i> 2009) or esotropia (Garg & Archer 2007, Stager <i>et al.</i> 2010).
<i>Visual acuity</i>	Amblyopia is uncommon.
<i>Cover test</i>	On the squinting day, a marked deviation for near and distance fixation is present. On the straight day, a latent deviation for near and distance fixation is present, which is usually exophoria.
<i>Binocular function</i>	Suppression is present on the squinting day with normal BSV present on the straight day. On a straight day, there is normal retinal correspondence with good fusion and stereopsis. On a squinting day, there is suppression but normal retinal correspondence is evident on correcting the angle of deviation in free space or using the synoptophore.

Management

Surgery is usually indicated for the manifest deviation (Stager *et al.* 2010). This may be performed once the deviation has become constant but may also be performed before the deviation has altered its cyclical pattern. As the deviation is often unilateral, a lateral rectus recession and medial rectus resection procedure on the deviating eye is usual. There is a good prognosis for restoration of BSV.

Non-specific exotropia

Exotropia that shows intermittent BSV, not conforming to any pattern.

Investigation

<i>Case history</i>	Symptoms of headache, asthenopia and intermittent diplopia may be appreciated by older patients.
<i>Visual acuity</i>	This is usually equal as the deviation is intermittent.
<i>Cover test</i>	The deviation is intermittently manifest at any fixation distance and there is no significant change in the angle of deviation for near or distance fixation.
<i>Binocular function</i>	Normal retinal correspondence but with weak control.
<i>Prism adaptation</i>	The angle of deviation is corrected in clinic using Fresnel prisms until there is no further increase in angle after a 2-hour period. This prism is then worn for a period of 2 weeks. If the deviation increases significantly, the surgery is planned for the increased angle. If the deviation is similar to the measured angle at the previous clinic visit, the surgery is planned for that measurement (Dadeye <i>et al.</i> 2003a).
<i>Differential diagnosis</i>	Decompensating exophoria

Management

These deviations often require surgery and may need post-operative treatment to consolidate the result. Surgery usually involves unilateral lateral rectus recession and medial rectus resection procedure.

Secondary exotropia

Exotropia that follows loss or impairment of vision.

Investigation

<i>Fundus and media</i>	Retinal changes relating to the cause of impaired vision may be evident. Causes include trauma, congenital loss of vision, retinal detachment, cataract, corneal pathology, glaucoma and aphakia (Dawson <i>et al.</i> 2005).
<i>Case history</i>	Loss of vision at birth or after the age of 7–8 years is often reported (Havertape <i>et al.</i> 2001).
<i>Cover test</i>	A moderate angle exotropia is present for all fixation distances.
<i>Visual acuity</i>	A severe visual deficit is observed and may be unilateral or bilateral.
<i>Ocular motility</i>	Limitations of ocular motility will be noted if there is associated damage to the globe.
<i>Binocular function</i>	This will be absent or reduced, dependent on the extent of visual loss.
<i>Suppression</i>	Post-operative diplopia risk must be evaluated in patients over 7 years of age. This may be done using prisms and under/overcorrecting the deviation. Botulinum toxin may also be used diagnostically (Khan <i>et al.</i> 2008). Intractable diplopia is rare but pre-operative assessment is essential (Kushner 2002).
<i>Angle of deviation</i>	The deviation is assessed by corneal reflections if there is poor fixation due to reduced visual acuity (Rowe 1994).

Management

Treat the cause of visual loss if possible. Surgery for removal of cataract often leads to spontaneous resolution of secondary exotropia (Chaudhuri & Pandey 2000).

Surgery: It is important to take into account the results of the post-operative diplopia test as the aim is for parallel visual axes or a slight overcorrection. For large angles greater than 50 prism dioptres, supramaximal resection/recession can be undertaken of up to 11 mm for medial rectus and 14 mm for lateral rectus (Chang *et al.* 2011). In general, a satisfactory result is obtained with surgery (Gusek-Schneider & Boss 2010).

Botulinum toxin: This is often a beneficial treatment with a low percentage of complications such as ptosis. Twenty percent are reported to continue injections and others proceed to surgery after botulinum toxin has evaluated the potential for binocular vision or diplopia with straighter alignment (Dawson *et al.* 2005).

Contact lens: If surgery is contraindicated due to the physical state of the eye, a painted contact lens may be used to mask the appearance of the eye.

Consecutive exotropia

Exotropia in a patient who has previously had esotropia or esophoria.

Investigation

<i>Case history</i>	The consecutive deviation may occur spontaneously with the passage of time but may also be iatrogenic in which there will be a history of prior surgical procedures for a convergent deviation. The change from esotropia to exotropia may occur from as young as 3 years of age (Senior <i>et al.</i> 2008). Symptoms of diplopia may be appreciated particularly immediately post-operatively and in older patients. Surgery with overcorrection may be deliberate, particularly for esotropia with an expected functional result. In cosmetic cases, overcorrection would be unplanned. Spontaneous cases occur in weak or absent fusion. There is a gradual onset with no symptoms and is commonly seen in older children and adults with a history of strabismus from early childhood and often associated with amblyopia (Ganesh <i>et al.</i> 2009).
<i>Visual acuity</i>	The patient is often amblyopic, particularly in cases of spontaneous unilateral exotropia.
<i>Cover test</i>	A manifest divergent deviation may be present either intermittently or constantly. The exotropia is usually seen first at distance fixation and then progressing to presence at near fixation also (Senior <i>et al.</i> 2008).
<i>Ocular motility</i>	Limitations may exist from previous strabismus surgery, e.g. lost muscle, maximum recessions.
<i>Binocular function</i>	Absent or weak binocular function will be demonstrated. Assess the potential for retinal correspondence particularly if the deviation was intermittent previously. The angle of deviation may be corrected in free space with prisms or on the synoptophore and the quality of binocular function evaluated.
<i>Angle of deviation</i>	Measure at near and distance fixation and with/without spectacle correction.
<i>Prism adaptation</i>	This is used to elicit the full angle of deviation to determine the need for further management and surgical intervention.

Management

When considering a corrective result, surgery is indicated if the appearance is poor with the aim for slight overcorrection.

When considering a functional result, treatment is indicated if the deviation has not resolved.

Concave lenses may be used in cases with a high AC/A ratio to induce accommodative convergence and thereby reduce the angle of divergent deviation.

Base-in prisms: The lowest strength needed to control the deviation is given and then decreased gradually over a period of time.

Orthoptic exercises to improve positive relative convergence and aid control of the deviation.

Botulinum toxin is useful for small angles to regain alignment and in cases at risk of post-operative diplopia (Lawson *et al.* 1998). It is particularly useful for those with the potential for binocular vision but of less benefit for those with absent binocular responses (Dawson *et al.* 1999).

Surgery should be considered for larger angles unresponsive to conventional therapy (Tejedor & Rodriguez 2007). The aim is for parallel visual axes as there is a good binocular prognosis.

Hypertropia

One eye is rotated upwards.

Hypotropia

One eye is rotated downwards.

Heavy eye phenomenon

This form of strabismus usually occurs in older children and adults.

Aetiology

This rare form of strabismus is due to a high degree of anisomyopia. The lateral rectus and superior rectus muscles are often displaced inferiorly and nasally, respectively (Krzizok & Schroeder 1999), and associated with restrictive abduction and elevation (Krzizok & Schroeder 2003). It may also occur in non-myopes and usually in elderly patients with age-related lateral rectus/superior rectus band degeneration that allows slippage of the muscles (Rutar & Demer 2009).

Investigation

<i>Visual acuity</i>	This may be reduced due to high refractive error.
<i>Cover test</i>	A hypo-deviation and esotropic deviation is typically seen in the primary position.
<i>Abnormal head</i>	There is often a head tilt to the side of the hypo eye (the posture more myopic eye).
<i>Ocular motility</i>	Abduction and elevation of the affected eye is usually seen.
<i>Binocular function</i>	BSV may be maintained by the use of the abnormal head posture and a good fusional range is noted. Binocular vision is absent with long-standing strabismus.

Management

The primary management option is surgical intervention and the aim is to realign the muscle paths. Loop myopexy is proposed in which a non-absorbable suture is looped around the lateral rectus and superior rectus to pull the muscles back into alignment (Durnian *et al.* 2010, Rowe & Noonan 2006). The muscle paths may also be realigned by muscle union (Yamaguchi *et al.* 2010).

Myopic strabismus

Aetiology

Bilateral high myopia causes displacement of the extraocular muscle paths within the orbit, which alters their primary and secondary rotator actions (Virdee 2007). Orbital imaging shows displacement of the rectus muscles. Typically, there is downward displacement of the lateral rectus muscles and nasal displacement of the superior and inferior rectus muscles (Oh *et al.* 2002, Clark *et al.* 1998). There is frequently greater displacement of the lateral rectus muscles with restricted abduction and elevation (Krizik & Schroeder 2003).

Investigation

<i>Refraction</i>	High bilateral myopia frequently in excess of -12.00 DS. Long axial length measurements obtained on biometry.
<i>Cover test</i>	Esotropia and hypotropia in primary position.
<i>Ocular motility</i>	Marked limitation of eye movements on abduction and elevation of either eye. A simulated Brown's syndrome pattern may be noted due to bilateral lateral rectus displacement inferiorly or heterotopy of the medial rectus muscles (Clark <i>et al.</i> 1998).
<i>Binocular function</i>	Presence of binocular vision is dependent on the duration of strabismus.

Management

Surgery is undertaken to realign the extraocular muscle paths. Muscle union of the superior and lateral rectus muscles is highly effective in restoring the dislocated globe back to the muscle cone (Durnian *et al.* 2010, Rowe & Noonan 2006, Yamaguchi *et al.* 2010).

Cyclotropia

One eye is wheel rotated so that the upper end of its vertical axis is nasal (incycloptropia) or temporal (excycloptropia).

Dissociated vertical deviation

When the vision of either eye is reduced, that eye deviates progressively upwards, but reverts to its original position when the embarrassment ceases. It may sometimes be so asymmetrical as to be virtually unilateral and may be associated with BSV or a manifest deviation. DVD is typically seen in the infantile esotropia syndrome. Early surgical intervention for infantile esotropia is reported to be associated with a reduced incidence of the development of DVD (Arslan *et al.* 2010).

Aetiology

The aetiology of DVD is obscure. Some theories include the following:

- DVD may be the result of altered interaction between cortical and subcortical pathways with a nasal bias in the ocular dominance columns of the primary visual cortex leading to asymmetric input to the omnipause vertical vergence system and cycloversion neurones in the interstitial nucleus of Cajal via the superior colliculus (ten Tusscher & van Rijn 2010).
- DVD may be considered to be a movement towards the position of absolute rest when fusion is disrupted.
- There may be a paretic element involving bilateral paresis of the depressor muscles.
- DVD may be due to a variety of associated factors including a return to the position of rest, relapse of peripheral fusion, unequal retinal stimulation and imbalance of the oblique muscles.
- Studies of monocular OKN have been found to be abnormal in normal infants up to a few months of age. The eyes respond to image movement from temporal to nasal but not from nasal to temporal. With increasing age, OKN becomes bidirectional. Asymmetry of monocular horizontal OKN persists in patients with strabismus associated with DVD. This asymmetry in monocular OKN is thought to be due to an abnormality of some of the temporal retinal fibres that are misdirected through the chiasm (Mein 1983).

Investigation

<i>Case history</i>	There is usually a history of manifest deviation from an early age. The parents may have noticed DVD when the patient looks to one side where it resembles an inferior oblique overaction. They may also notice spontaneous elevation (Fig. 6.8) that tends to occur if the patient is daydreaming or is excessively fatigued. An abnormal head posture is present in some cases with tilt usually to the side of the fixing eye (Bechtel <i>et al.</i> 1996, Crone 1954, Santiago & Rosenbaum 1998). Symptoms are rare.
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	DVD is part of the infantile squint syndrome where patients have infantile esotropia with manifest nystagmus. The nystagmus is manifest latent in type. DVD is usually demonstrated around the age of 2 years (Arslan <i>et al.</i> 2010).
<i>Visual acuity</i>	Amblyopia may be present if there is a unilateral manifest deviation. Vision may be reduced unioocularly compared to binocularly due to manifest latent nystagmus.
<i>Cover test</i>	A manifest deviation is typically present that is often esotropic. Nystagmus is usually horizontal, but may have a vertical or rotational component. Progressive elevation of either eye is seen under cover and a slow downward movement when the cover is removed. The upward movement of the eye is slow, more marked on distance fixation and on prolonged dissociation. When the occluder is removed, the eye drifts down to the midline and occasionally may move below the midline before moving up again to refixate. DVD is often more marked on distance fixation.
	Rarely, dissociated hypotropia may be noted on cover test, which is an uncommon variant of DVD (Kraft <i>et al.</i> 2006).
<i>Ocular motility</i>	DVD is a bilateral condition but may be asymmetric (von Noorden 1988a,b). There may be apparent or true inferior oblique overactions (Helveston 1969, McCall 1991). In cases with inferior oblique overactions, a V eso pattern is expected.
<i>Binocular function</i>	Absent or defective fusion is detected. Normal BSV is rare.
<i>Monocular OKN testing</i>	This is useful in young children with infantile esotropia in whom testing DVD is suspected but not confirmed on cover test. A defective nasal to temporal response is expected (Fitzgerald & Billson 1984).
<i>Differential diagnosis</i>	

	DVD	Inferior oblique overaction
<i>Cover test</i>	Hyper-deviation remains the same in primary position and contralateral versions	Hyper-deviation increases on contralateral versions
<i>Ocular motility</i>	Sudden upshoot on contralateral versions when nose intervenes	Gradual updrift on contralateral versions
<i>A/V pattern Bielschowsky phenomenon</i>	Mainly A pattern Positive	V pattern Negative

<i>Dark wedge test</i>	This is also known as the Bielschowsky phenomenon and differentiates between DVD and alternating hyperphoria. This is described in Chapter 3.
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Figure 6.8 Hypertropia.

Measurement of angle Prism cover test measurements should be made for the vertical DVD angle alongside any associated horizontal angle. In addition, a +6 DS lens may be used instead of a standard occlude. This allows visualisation of the eye and the lens strength is high enough to blur the image for the patient but not eliminate the image formation on the retina (Dadeye *et al.* 2003b).

Management

Correct the refractive error and treat amblyopia if present. Manifest strabismus is treated according to the type and size of deviation.

DVD is managed surgically when indicated by spontaneous elevation, which is cosmetically unacceptable. Surgery may involve a recession or a faden procedure to the superior recti muscles (Esswein *et al.* 1992). The amount of recession may be varied when asymmetrical. A faden procedure may be combined with superior recti recessions if there is a significant vertical deviation associated with the spontaneous elevation (Duncan & von Noorden 1984). Anterior transposition of the inferior oblique may also be considered and shows significantly reduced amounts of DVD whether associated with inferior oblique muscle overaction or not (Black 1997, Burke *et al.* 1993, Kratz *et al.* 1989, Nabie *et al.* 2007, Ruttum 1997, Seawright & Gole 1996, Strominger *et al.* 2009, Varn *et al.* 1996). Surgery for residual deviations may involve inferior rectus resection as a second procedure (Esswein Kapp & von Noorden 1994).

Where DVD is associated with inferior oblique overaction, a combined inferior oblique resection and anterior transposition can be beneficial (Farvardin & Attarzadeh 2002, Lehman *et al.* 2005). Dissociated hypotropia responds well to large inferior rectus recessions (Kraft *et al.* 2010).

Dissociated horizontal deviation

DHD is reported as a clinical expression of dissociated esotropia with a common clinical presentation as an intermittent exo-deviation of one eye resulting from the

superimposition of a dissociated eso-deviation on a baseline exo-deviation (Brodsky & Fray 2007).

Investigation

<i>Case history</i>	There is frequently a history of previous strabismus surgery (Gamio 2011).
<i>Visual acuity</i>	Amblyopia is common.
<i>Cover test</i>	There is intermittent asymmetrical abduction and elevation of the dissociated eye (Wilson & McClatchey 1991, Zubcov <i>et al.</i> 1990). Associated DVD is common and patients may also have associated vertical element, and there may be latent nystagmus and ex-cyclotropia (Wilson <i>et al.</i> 1995). The exo-deviation is smaller with fixation of the non-preferred eye than fixation with the preferred eye (Brodsky 2007).
<i>Sbiza bar</i>	The deviation decreases with neutral density filters (Wilson & McClatchey 1991).
<i>Reversed fixation</i>	The eye movements are observed under a prism cover test during movement of the occluder. Where movement of the occluded eye occurs after neutralisation of the angle has been achieved, this is said to indicate presence of DHD (Brodsky <i>et al.</i> 2005).
<i>Angle of deviation</i>	This should be measured fixing either eye to document discrepancy of angle.

Management

Surgical treatment, if required, is with lateral rectus recessions (Wilson *et al.* 2000) and may be combined with superior rectus recession if there is an associated vertical element or DVD (Gamio 2011).

Quality of life

The presence of strabismus, whether with the potential for binocular vision or not, can have a detrimental effect on quality of life leading to psychosocial issues. A number of quality of life instruments (questionnaires) are available to assess issues for children and adults with strabismus, such as the AS-20, which is a 20-item adult strabismus questionnaire, the VFQ25 questionnaire, which is a visual function questionnaire and the DAS59 questionnaire, which is the Derriford Appearance Scale (Durnian *et al.* 2009, Hatt *et al.* 2009, Leske *et al.* 2010). The Amblyopia and Strabismus Questionnaire (A&SQ) is designed specifically to assess quality of life in amblyopia and/or strabismic patients. This questionnaire has shown that visual acuity of the amblyopic eye correlates with all five of the A&SQ domains (van de Graaf *et al.* 2007).

Key questions of quality of life measurements include effects of vision on self care, mobility, participation in work and leisure activities, interaction with other people and sense of oneself (Misajon *et al.* 2005).

Pseudostrabismus

This is an appearance of strabismus when no manifest deviation of the visual axes is actually present. These patients require full investigation as the prevalence of strabismus is higher in those diagnosed with pseudostrabismus than for the general population and particularly for those with developmental delay (Pritchard & Ellis 2007).

Aetiology

1. Epicanthus may give a convergent appearance where skin folds are prominent between the medial canthi and the nose (Fig. 6.9). Lateral canthus may give a divergent appearance but this is relatively rare.
2. Angle kappa is the angle between the pupillary axis and the visual axis. The corneal reflections may appear slightly nasal producing a positive angle kappa. A large angle kappa gives an appearance of a divergent deviation. If both axes coincide, corneal reflections appear right in the centre of the pupil producing a negative angle kappa giving an appearance of a convergent deviation. Synoptophore slides may be used to measure the degree of angle kappa (see Chapter 3).
3. Inter-pupillary distance. A wide IPD gives the appearance of a divergent deviation. A narrow IPD gives the appearance of a convergent deviation.
4. Enophthalmos gives the appearance of a convergent deviation and exophthalmos, a divergent deviation.
5. Ptosis may give appearance of a vertical deviation.
6. Asymmetry of palpebral fissure.
7. Facial asymmetry.
8. Ill-fitting spectacles.
9. Presence of an abnormal head posture.
10. Heterochromia iridum – a difference in the iris colour of either eye.
11. Iris coloboma – a fissure or gap in the iris.
12. Anisocoria – a difference in the pupil size of either eye.



Figure 6.9 Pseudostrabismus.

Management

Orthoptic investigation will establish the presence of BSV and fusional control in cases of pseudo-strabismus. However, if it is found to be associated with true strabismus, further treatment should be given as indicated.

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7

Microtropia

Microtropia is a small-angle heterotropia (usually of ≤ 10 prism dioptres) in which a form of binocular single vision occurs. When there is an associated heterophoria it is known as microtropia with a latent component.

The incidence of microtropia is 2–3% of which most cases are microesotropia with microexotropia occurring less commonly.

Terminology

Fixation disparity, fusion disparity, convergence fixation disparity, pathologic fixation disparity and fixation disparity with neuroanomalous correspondence are terms that have been used to describe a physiological disparity occurring in Panum's areas in cases of heterophoria with foveal suppression. Jampolsky (1951, 1956) commented on the presence of central suppression and peripheral fusion in small angle deviations. The central suppression prevents diplopia of the fixation object. Peripheral fusion with normal retinal correspondence is achieved, as Panum's fusional space is large enough to compensate for the small angle deviation.

Eso flick, flicker cases and ultra-small angles are terms that have been used to describe cases of minimal manifest deviation detectable on cover test (Bryer 1953).

Parks & Eustis (1961) used the term monofixation phoria to describe deviations < 8 prism dioptres, which exhibited characteristics of a phoria and a tropia and which might occur primarily due to an inability to fuse macular images, secondary to treatment of large-angle strabismus, secondary to anisometropia or secondary to a unilateral macular lesion. Peripheral fusion with normal retinal correspondence in the presence of a foveal suppression scotoma was thought possible due to a stretched Panum's area (Jampolsky 1956). Bagolini glasses and after-image testing suggested that abnormal retinal correspondence was present rather than normal. Bagolini & Capobianco (1965) demonstrated an enlarged Panum's area with abnormal retinal correspondence.

The terms microstrabismus and microtropia were introduced by Lang (1969a, 1969b). This emphasises the manifest part of the condition and is therefore

considered to be most appropriate. The term describes cases of small-angle tropia with harmonious abnormal retinal correspondence, gross stereopsis and slight amblyopia. Helveston & von Noorden (1967) used the term to describe a condition with amblyopia, suppression scotoma, abnormal retinal correspondence, an absence of movement on cover test and parafoveal eccentric fixation (microtropia with identity).

In addition to the term microtropia, the use of the term subnormal binocular vision was used by von Noorden as a category to describe binocular cooperation. Subnormal binocular vision included orthotropia or heterophoria, normal visual acuity in both eyes, fusional amplitudes, normal retinal correspondence, foveal suppression in one eye in binocular vision and reduced or absent stereopsis.

Most authors now encompass the term microtropia for both positive and negative cover test findings (with and without identity).

Classification

Microtropia is usually associated with abnormal retinal correspondence (Lang 1969) but also with normal retinal correspondence with an expanded Panum's fusional area (Gittoes-Davies 1951, Jampolsky 1956). Microtropia may be:

- *with identity*, which occurs when there is harmonious abnormal retinal correspondence and absolute eccentric fixation, and the angle of anomaly equals the angle of eccentricity. A manifest deviation is not seen on cover test;
- *without identity*, which may have central or non-absolute eccentric fixation and the retinal correspondence may be:
 - abnormal retinal correspondence
 - normal retinal correspondence with central suppression and peripheral fusion.

A manifest deviation will be seen on cover test.

Microtropia may be further classified as:

- *Primary microtropia*: There is no history of previous large-angle strabismus. Microtropia is usually stable and deterioration into a large angle is not common (Arthur *et al.* 1989).
- *Primary decompensating*: These cases have an accommodative element with hypermetropia. Onset is between 1–3 years usually where the deviation has decompensated into a larger angle.
- *Secondary microtropia*: This follows optical or surgical reduction of a primary concomitant larger angle deviation, for example fully accommodative esotropia reduced to microtropia.

Aetiology

A number of aetiological factors are associated with microtropia, the most common being anisometropia but also foveal pathology, hereditary factors and the presence of a congenital defect.

Anisometropia

The high incidence of anisometropia suggests a possible aetiological relationship (Helveston & von Noorden 1967, Setayesh *et al.* 1978). Microtropia may develop secondary to a foveal scotoma caused by uncorrected anisometropia during early infancy (Helveston & von Noorden 1967). It is proposed that uncorrected anisometropia presents a blurred image to the eye with the higher refractive error and a foveal suppression scotoma develops with subsequent development of parafoveal fixation and microtropia.

This may lead to eccentric fixation under monocular conditions and abnormal retinal correspondence under binocular conditions (von Noorden 1970). Anisometropia may decrease with age and therefore the refractive error may not be evident on examination of the child at later presentation to the clinic.

Foveal pathology

Microtropia resulting from *foveal pathology* may include cases of infection with scar formation (e.g. toxocara) or foveal haemorrhage at an early age.

Congenital

There may be an inherent inability or loss of a prior ability for bifoveal fusion (Gittoes-Davies 1951, Parks 1974, Wilson *et al.* 1993). There may also be a variant in the fixation reflex feedback mechanism. Precision in the development of the fixation reflex is essential for the evolution of normal binocular single vision and an anomaly of this reflex has been postulated as an aetiological factor in microtropia and eccentric fixation (von Noorden 1970). A variation in the interaction between the feedback of uniocular fixation and the feedback of binocular fixation may exist.

Hereditary factors

There is a high incidence of associated anomalies of binocular vision in families of microtropic patients, which suggests that it may be caused by multifactorial inheritance of refractive errors or motor and sensory anomalies of binocular vision (Cantolino & von Noorden 1969).

Investigation

Refraction

Patients are often anisometropic, which has been reported in up to 80% of cases. The amblyopic eye is commonly the more hypermetropic with or without astigmatism. With microtropia exo, anisometropia is usually of mixed astigmatism. With microtropia eso, anisometropia is usually spherical. If due to a macular lesion, this may be detected on fundoscopy.

Fundus and media

The fovea and fundus should be examined for presence of central retinal abnormalities (Salvi *et al.* 2007).

- Case history* Primary microtropes often present later with referral from school vision checks. Primary decompensating microtropes may present with symptoms if the latent component becomes decompensated. The previous treatment is particularly important and may have included optical correction, orthoptic treatment or surgery for a large deviation, anisometropia or an accommodative esotropia.
- Visual acuity* Varying degrees of amblyopia of the deviating eye may be noted. The crowding phenomenon is a characteristic of microtropia. Since the fovea of the deviating eye is suppressed the beginning of words may be blurred or absent in left microesotropia and the end of words blurred or absent in right microtropia. This is important to consider when testing linear visual acuity and may occasionally be symptom producing with difficulties in reading text. Separation difficulties tend to be more notable on near fixation.
- Cover test* With identity there is no manifest deviation, but a latent deviation may be present. The size of the very small angle of deviation equals the distance in the amblyopic eye between the fovea and the area used for eccentric fixation. Because of eccentric fixation, the parafoveal area of the eye is used for binocular as well as monocular vision. Therefore, no refixation movement is required by the amblyopic eye when the foveally fixing eye is covered.
- Without identity, a slight manifest deviation is detected. There may also be an associated latent component that is usually but not invariably in the direction of the heterotropia. This will be detected by alternate cover test. Where there is an associated heterophoria, the recovery is to the microtropia rather than the bifoveal fixation. The rate of recovery is very important since decompensation of the heterophoria may become symptom producing.
- Vertical microtropia is rare but is sometimes seen in patients with congenital or long-standing vertical muscle palsies.
- Bagolini glasses* A central scotoma may be detected. Under normal circumstances, a symmetrical cross is expected. A central or paracentral gap in the line seen by the affected eye is reported by some patients.
- 4 dioptre prism test* This is assessed base out and base in for microtropia with identity to differentiate from bifoveal fixation with heterophoria. If a base-out prism is placed before the unaffected eye, it will make a movement inwards to overcome the effect of the prism. By Hering's law, the other eye will also move conjugately by the same amount. However, as the amount of movement is so small, the deviated image remains within the scotoma area of the affected eye and no disjunctive recovery movement of the affected eye will be seen. When the base-out prism is placed before the affected eye, there will be no movement of that eye as the image remains within the scotoma area, and consequently, no associated movement of the other eye.

The base-in prism is used particularly for microextropia (Epstein & Tredici 1972).

Fixation

This should be assessed with a viscuscope, particularly if no deviation is seen on cover test.

Eccentric fixation will be seen in microtropia with and without identity with absolute and non-absolute eccentricity, respectively. Eccentric fixation in microtropia occurs with sensory motor adaptation of fixation at the edge of the foveal scotoma. Fixation is typically parafoveal or parafoveolar. In microesotropia, the fixation is nasal and slightly superior to the fovea. In microexotropia, fixation is temporal and/or superior to the fovea (Johnson *et al.* 1981).

Binocular function

Microtropia with identity is associated with absolute eccentric fixation with abnormal retinal correspondence. Microtropia without identity may demonstrate harmonious abnormal retinal correspondence but some cases may have central suppression and peripheral fusion and normal retinal correspondence, particularly those with a flick and associated phoria. Cases of harmonious abnormal retinal correspondence that show an associated latent component do so because the abnormal retinal correspondence is so well developed that the latent element develops around the fixation point.

A good range of peripheral fusion may be demonstrated and the patient often appreciates diplopia when fusion fails.

Reduced stereoacluity is present but to a varying degree depending on the level of visual acuity. Random dot tests are reported as being more difficult to perceive and some patients show absent stereoacluity (Matsuo *et al.* 2003).

In microtropia, suppression only involves the fovea and the diplopic and fixation point of the deviating eye.

Angle of deviation

An alternate prism cover test will measure the total deviation. If an associated latent deviation is present, a simultaneous prism cover test will measure only the manifest portion of the deviation. This measurement must be <10 prism dioptres for a diagnosis of microtropia. The difference between the alternate prism cover test and simultaneous prism cover test is the amount of heterophoria kept latent by peripheral fusion. When performing, the simultaneous prism cover test, the prisms are placed before the microtropic eye and a cover/uncover test used on the other eye until the movement of the affected eye is neutralised.

Synoptophore

In cases of central fixation, the synoptophore can be used to find an angle of anomaly, so proving the presence of abnormal retinal correspondence. In practice, this is very difficult to demonstrate, as the angle of deviation is so small. Foveal slides are used. There will be no angle of anomaly in cases of normal retinal correspondence.

Management

Correct the refractive error and treat amblyopia to achieve optimum visual acuity. If there is low visual acuity, more intensive occlusion may be recommended depending on age. Otherwise, part-time occlusion is suitable. Occlusion is continued until there is no further improvement in acuity. While occlusion treatment is undertaken, the orthoptic investigation must include assessment of fixation, suppression and fusional vergence to ensure adequate control of the deviation and guard against possibility of intractable diplopia. The risk of intractable diplopia has been deemed a contraindication to treatment in some cases (Lang 1969a, 1969b). However, this is unlikely given the presence of good peripheral binocular function in many cases. Decompensation of the angle of deviation is unlikely as patients usually have comfortable and near-normal binocular function with good peripheral fusion.

There are reports of poor acuity levels in microtropia despite treatment (Evard-Halm & Mailette de Buy Wenniger-Pick 1989) and it has been suggested that, with treatment, vision does not improve beyond 6/9 and there is at least one line difference in acuity between both eyes (Lithander & Sjostrand 1991). Amblyopia treatment should continue as long as visual acuity is improving and stopped only when improvement in acuity ceases. It has been thought that the sensory status is irreversible. However, there are reports of elimination of microtropia and amblyopia following occlusion treatment (Cleary *et al.* 1998, Houston *et al.* 1998, Henshall & Rowe 1999). Fixation becomes foveal with improved stereoacuity and resolution of the microtropia.

It is usual to treat the microtropia as if there is normal single vision. Therefore, any associated heterophoria that seems to be decompensating can be treated with orthoptic exercises to improve the fusional range. Negative relative convergence should be improved for microesotropia and positive relative convergence for microexotropia. In addition, bar reading, prisms and stereograms can be utilised. Prisms may be long-term and incorporated into spectacle prescription for small angles. Botulinum toxin has been shown to be beneficial for the management of small-angle decompensating microtropia (Ripley & Rowe 2007, Bansal *et al.* 2008). Surgery may be undertaken if there is a large angle.

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8 Amblyopia and Visual Impairment

Amblyopia is a condition of diminished visual form sense which is not as a result of any clinically demonstrable anomaly of the visual pathway and which is not relieved by the elimination of any defect that constitutes a dioptic obstacle to the formation of the foveal image. It occurs in up to 4% of the population (Loudon & Simonsz 2005).

Classification

- *Stimulus deprivation amblyopia*: Amblyopia, which is the result of lack of adequate visual stimulus in early life. This may be unilateral or bilateral and may be:
 - complete, where no light enters the eye
 - partial, where there is some passage of light into the eye.
- *Strabismic amblyopia*: Amblyopia, which is the result of manifest strabismus and is caused by constant unilateral strabismus in childhood.
- *Anisometropic amblyopia*: Amblyopia, which is the result of a significant difference in the refractive errors of the two eyes where one eye has the visual advantage at all distances.
- *Meridional amblyopia*: Amblyopia, which is the result of uncorrected astigmatism where one or both eyes are predominantly astigmatic.
- *Ametropic amblyopia*: Bilateral amblyopia, which is the result of a high degree of uncorrected bilateral refractive error.
- *Occlusion amblyopia*: Amblyopia, which may occur after use of total occlusion or atropine, particularly before the age of 2 years. Visual acuity is usually restored with careful treatment and monitoring.

Aetiology

Amblyopia is caused by inadequate stimulation of the visual system during the critical period of visual development in early childhood (<6 years) (Daw 1998). This is most marked under the age of 2 years (Awaya *et al.* 1987). The site of amblyopia is

proposed as the visual cortex (area V1) where there are marked changes with loss of binocular driven cells and loss of neurones driven by the amblyopic eye (Kiorpes & McKee 1999, Kozma & Kiorpes 2003). Changes are also found in area V2.

Amblyopia may be unilateral or bilateral and the cause may be any or a combination of the following factors:

- *Light deprivation:* There is no stimulus to the retina. This is uncommon as it is likely that some light enters the eye even in dense cataract (white noise).
- *Form deprivation:* The retina receives a defocused image as with refractive errors.
- *Abnormal binocular interaction:* Non-fusible images fall on each fovea as with strabismus.

The prognosis for achieving good visual acuity decreases when more than one of these factors is present together in one case.

There has been a decrease in amblyopia over recent years that may be due to better screening and detection (Kvarnstrom 1998, Latvala 1996).

Investigation

Refraction Anisometropia, high astigmatism and bilateral high refractive errors are aetiological factors.

Case history Early onset unilateral, constant strabismus with long duration without treatment predisposes to strabismic amblyopia.

Congenital ocular abnormalities such as cataract and ptosis, predispose to stimulus deprivation amblyopia (Anderson & Baumgartner 1980).

Visual acuity Many aspects of visual function are adversely affected by the presence of amblyopia including high contrast linear, single optotype, repeat letter and low-contrast linear, vernier and displacement thresholds (Simmers *et al.* 1999).

If possible, it is better to use a linear test type: logMAR is considered gold standard (Cleary 2007). The patient may demonstrate the crowding phenomenon where reduced visual acuity is detected on linear testing despite better visual acuity on single optotype testing. This is due to additional stimulation from contours of adjacent letters in a linear test type and a reduction in lateral retinal inhibition that produces confusion (Hess *et al.* 2000). Assessment of fixation aids determination of visual acuity. Free alternation indicates equal visual acuity. Holding fixation beyond a blink indicates good visual acuity. Strong objection to occlusion of the good eye and no objection to occlusion of the deviating eye suggest severely reduced visual acuity. This is also indicated with searching eye movements of the deviating eye.

	Visual acuity tends to be better at near rather than distance fixation (von Noorden & Frank 1976).
<i>Contrast sensitivity</i>	Reduced thresholds at different spatial frequencies are seen in amblyopia. Stimulus deprivation amblyopia may cause defects at high spatial frequencies whereas defects in all spatial frequencies are seen with combined strabismic and anisometropic amblyopia. Contrast sensitivity is not correlated with visual acuity. Contrast sensitivity is also reduced in the fellow eye (Chatzistefanou <i>et al.</i> 2005).
<i>Cover test</i>	There may be a unilateral constant strabismus. By observing fixation and reaction to occlusion, an estimate of visual acuity can be obtained as detailed above. However, binocular fixation preference is not reliable for detection of strabismic amblyopia (Procianoy & Procianoy 2010) and strong fixation behaviour may be more representative of strong ocular dominance (Hakim 2007). There may be ptosis that can be unilateral or bilateral. Congenital ptosis is associated with amblyopia of strabismic, anisometropic and stimulus deprivation types (Dray & Leibovitch 2002).
<i>Visuscope</i>	Fixation may be eccentric or wandering, in a parafoveal, macular or peripheral retinal area. Generally, the further from the fovea, the poorer the visual acuity and the poorer the prognosis for management of amblyopia.
<i>Neutral density filter</i> (Fig. 8.1)	This test makes use of the fact that amblyopic eyes perform relatively well under conditions of low illumination (scotopic state). The visual acuity when tested with the filter is not significantly reduced in the amblyopic eye when compared with visual acuity of the normal eye. This test differentiates between amblyopia and reduced visual acuity due to organic defects, as organic amblyopia will have further reduction in visual acuity under poor illumination (Burian 1967). For example, neutral filters profoundly reduce vision in eyes with central retinal lesions and glaucoma, whereas the vision of eyes with amblyopia is not reduced with filters (von Noorden & Burian 1959, Sloan 1969, Hess & Howell 1978, France 1984).
<i>Binocular vision</i>	Fine motor skills performances of children with amblyopia are poor and, particularly, so for strabismic amblyopia (Webber <i>et al.</i> 2008). Presence of binocular vision is important for eye-hand coordination (Suttle <i>et al.</i> 2011).

Management

Correct the refractive error as visual acuity often responds when the correct prescription is worn (refractive adaptation). A period of 18 weeks is recommended for refractive adaptation before implementation of occlusion (Cleary 2007, Shotton

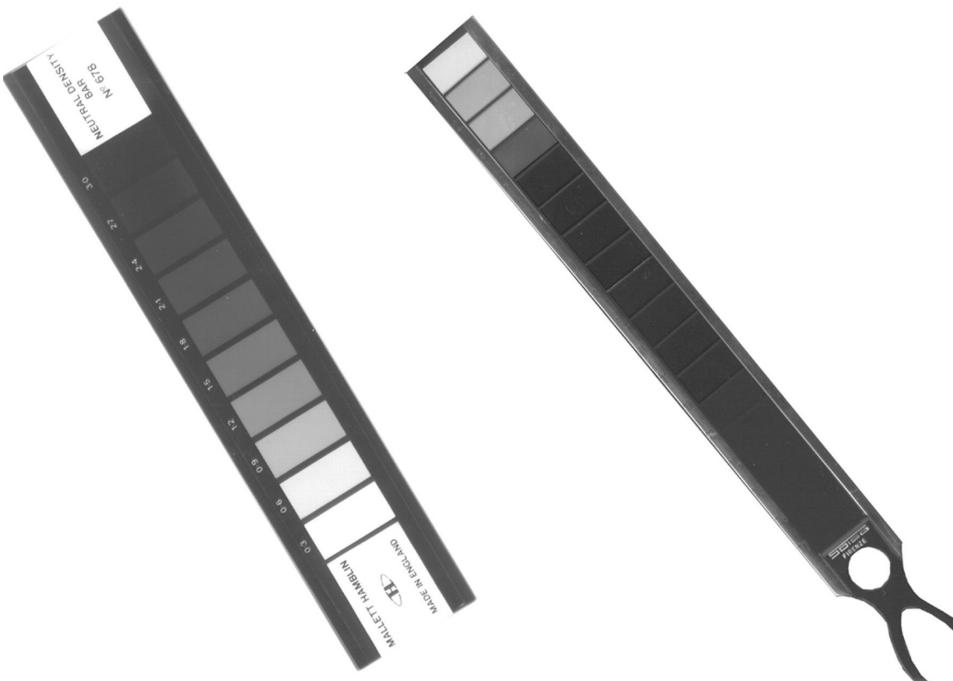


Figure 8.1 Neutral density filter bar.

et al. 2008, Moseley *et al.* 2009). A mean improvement of 0.24 log visual acuity with spectacle prescription only has been reported with a mean time to best visual acuity of 14 weeks (maximum 18 weeks) (Moseley *et al.* 2002, Stewart *et al.* 2004b). Roughly, one quarter to half of patients did not require occlusion after refractive adaptation (Stewart *et al.* 2004a, Chen *et al.* 2007). Refractive correction is particularly successful for children with untreated anisometropic amblyopia (PEDIG 2006).

Photorefractive keratectomy has been used for treatment of anisometropic amblyopia showing significant improvements in refractive error and visual acuity (Paysse *et al.* 2006).

Occlusion is the most commonly used method of treating amblyopia. The normal eye is occluded and occlusion may be in the form of total light or form, or partial. Clear explanation and instructions must be provided for parents, patients and care-givers:

- *Total light and form:* Opticlude and Coverlet are most widely used. Elastoplast is also available.
- *Total form:* Blenderm, Micropore, Frosted lens and Bangerter foils.
- *Partial:* Fablon and Sellotape.

Duration of occlusion

The starting level of occlusion is between 2 and 6 hours daily coupled with near or distance activities. Two hours of occlusion daily has been shown to be as effective as 6 hours daily for amblyopia of 0.3–0.6 logMAR in children under the age of 7 years (PEDIG 2003, PEDIG 2006). For those with amblyopia of 0.7–1.3, 6 hours of occlusion is as effective as full-time (Holmes *et al.* 2003, Stewart *et al.* 2007b). Approximately 50% fail to improve beyond 0.4 logMAR (PEDIG 2003, Stewart *et al.* 2007a) and approximately 50% achieve visual acuity of 6/9 or better in the treated eye (Woodruff *et al.* 1994, PEDIG 2002).

An ideal goal is less than 400 hours of occlusion or 6 months of occlusion treatment. The bulk of treatment effect is achieved with 150–250 hours of occlusion with a dose response of approximately 1 log line/120 hours of occlusion (Stewart *et al.* 2005, 2007b). Dose response does not appear related to type of amblyopia. However, it takes longer to achieve the same result with older children (Stewart *et al.* 2005). Late occlusion (after the age of 8 years) has been shown to be effective in selected cases (Mohan *et al.* 2004, PEDIG 2005). In the age group of 7–12 years, significant improvements in visual acuity have been documented in 53% with improvement in 25% for those aged 13–17 years (Mohan *et al.* 2004, Park *et al.* 2004, PEDIG 2005). Visual acuity results are better when patients have never been treated for amblyopia previously.

Risks of occlusion

1. Intractable diplopia may occur in patients with strabismus and should be carefully monitored if giving occlusion after the age of 6 or 7 years due to the risk of overcoming their suppression. Patients with anisometropic amblyopia usually have binocular single vision and therefore occlusion after the age of 8 years is possible but binocular function must be monitored carefully to ensure loss of good control of a latent deviation does not occur.
2. Occlusion amblyopia.
3. Dissociation of latent/intermittent deviation.
4. An increase in angle of deviation may occur on removal of occlusion.
5. DVD: The eye may elevate further.
6. Allergic response.
7. Infection may be incubated by the patch.
8. Danger socially due to disorientation.
9. Increased likelihood of bullying and stigmatisation (Horwood *et al.* 2005, Koklanis *et al.* 2006).

Consequences of amblyopia

The risk of permanent visual loss in the better eye is reported as 32.9 per 100,000 population (Rahi *et al.* 2002). Presence of amblyopia interferes with schooling, work, lifestyle, sports and career choice (Adams & Karas 1999, Packwood *et al.*

1999) because of poor vision, related impairment of motor tasks and binocular vision (Gray & Regan 1998) and deficits of prehension including reaching and grasping of objects (Grant *et al.* 2007).

Aims of occlusion

1. Equalise visual acuity
2. Achieve optimum visual acuity
3. Central fixation

Continue occlusion until:

1. equal visual acuity is achieved;
2. the optimum visual acuity is achieved;
3. there is no further increase in visual acuity with full-time total occlusion.

Recurrence

Long-term follow-up of amblyopic patients shows an average reduction in visual acuity for up to 75% of patients of 1.2–2.6 Snellen lines at least 5 years post cessation of treatment (Gregersen & Rindziunski 1965, Sparrow & Flynn 1979, PEDIG 2004, Bhola *et al.* 2006, King *et al.* 2007, De Weger *et al.* 2010) with a 1–4 line reduction for strabismic amblyopia and 3 line reduction for anisometropic amblyopia. Regression of visual acuity is more likely to occur with abrupt cessation of occlusion rather than tapered (Holmes *et al.* 2004). Risk of recurrence is reported as inversely correlated with age (Bhola *et al.* 2006). Mixed amblyopia may be more likely to recur than pure anisometropic or strabismic amblyopia (Tacagni *et al.* 2007). Other factors associated with deterioration of visual acuity are visual acuity of the amblyopic eye at the start of treatment and esotropia of $>4^\circ$. Visual acuity recovers with prompt re-treatment and good compliance (DeWeger *et al.* 2010). A follow-up period of 24 months is recommended following cessation of treatment of amblyopia in order to detect recurrence.

For older children treated for amblyopia (aged 7–12 years), visual acuity was sustained in most children for at least 1 year after cessation of amblyopia treatment (PEDIG 2007).

Compliance issues

Success of occlusion treatment relates in part to the compliance of the patient and parents/guardian in undertaking the occlusion regime. It is generally agreed that thorough discussion of the occlusion regime backed up by written information has a positive impact on occlusion success (Newsham 2002). Compliance has been reported in 78% in those with written information and 57% in those without information (Loudon *et al.* 2006). Compliance is noted to be better when visual

acuity is seen to improve and the child is not thought to be suffering socially or educationally (Dixon-Woods *et al.* 2006).

The age of patient, type of amblyopia, type of occlusion, level of acuity, length of time it must be worn each day and length of time period of the occlusion treatment also play a part in the general compliance and ultimate outcome of occlusion treatment.

Use of dose monitors and diaries of occlusion wear have been used to monitor occlusion compliance. Average compliance is reported in up to 54% of cases (Newsham 2000, Stewart *et al.* 2004b). Occlusion dose monitor compliance has been reported as 80% in those with satisfactory visual acuity improvement compared to 34% in those with unsatisfactory improvement (Loudon *et al.* 2003).

Factors influencing outcome include the presence of anisometropia, visual acuity at the start of treatment and compliance with treatment. Poor acuity or fixation at the start of treatment is a significant factor in the outcome (Beardsell *et al.* 1999).

Unresponsive amblyopia

Where visual acuity does not improve with occlusion treatment, an increase in occlusion or switch to another treatment option should be considered. Where continued lack of improvement occurs, detailed assessment of the visual pathway should be made to exclude pathology.

Cycloplegic drugs

Where occlusion is not tolerated, cycloplegic drugs may be used to blur the vision in the better eye, thereby giving the amblyopic eye more stimulus. Typically, atropine 1% has been used once daily but atropine instilled only at weekends is equally effective (PEDIG 2004, Repka *et al.* 2009). Atropine has been shown to be equally as effective as occlusion for visual acuity of 0.3–0.7 logMAR (PEDIG 2002) with maintenance of visual acuity to long-term follow-up (PEDIG 2008).

Advantages

1. The patient cannot cheat as can occur with occlusion where the child peeps over the patch or where there is poor or non-compliance.
2. The child and parent often prefer it to occlusion.
3. There is little or no cosmetic problem.

Disadvantages

1. Side effects of atropine.
2. Visual acuity may not be reduced enough where there is dense amblyopia.
3. Atropine takes a period of 10–14 days to wear off.
4. Frequent visits are required to monitor fixation as an indicator of visual acuity.

Penalisation

This is a further option where occlusion treatment is not tolerated. Penalisation is the treatment of amblyopia by optical reduction of form vision of the non-amblyopic eye at one or all fixation distances. The effect may be achieved by the alteration of the spectacle correction or use of Bangerter foils and may be enhanced by the use of a cycloplegic drug (Gregersen *et al.* 1965, Repka & Ray 1993).

Total penalisation is the use of the amblyopic eye at all distances by using strong convex lenses before the better eye. Near penalisation is the use of the amblyopic eye for near fixation by using a cycloplegic drug in the better eye and adding a convex lens up to 3.0 DS before the amblyopic eye. Distance penalisation is the use of the amblyopic eye for distance fixation by using additional convex lenses before the better eye. One trial has shown that the augmentation of weekend atropine treatment with a plano lens does not result in any significant differences when compared to weekend atropine treatment alone (PEDIG 2009).

Bangerter foils may be used as a form of optical penalisation (PEDIG 2010) but the optical characteristics of the foils do not correlate with their labelled density designation (Perez *et al.* 2010).

Computerised treatment

Interactive computerised systems are showing promise for treatment of amblyopia (Eastgate *et al.* 2006, Waddingham *et al.* 2006). Virtual reality based display systems have been developed in which both eyes are stimulated simultaneously [interactive binocular treatment (I-BiT) system]. The system incorporates two-dimensional and three-dimensional games and videos via a stereoscopic binocular display. There is potential for use of this system particularly for those non-compliant with conventional occlusion.

Drugs

Levodopa and Carbidopa have been used to improve visual acuity often in conjunction with occlusion therapy (Nahata *et al.* 2000, Mohan *et al.* 2001). These drugs are neurotransmitters that increase dopamine levels. The improvement is believed due to the general role of dopamine both in the retina and visual pathway (Gottlob & Strangler-Zuschrott 1990, Harris *et al.* 1990, Gottlob *et al.* 1992, Leguire *et al.* 1993, Leguire *et al.* 1995).

An improvement in vision is dose dependent with a higher dose showing a better response. Patients with very poor starting vision or sensory deprivation amblyopia respond poorly to treatment. Some reduction in vision is generally noted after treatment is stopped (Pandey *et al.* 2002). Treatment appears to be more effective in improving visual acuity in patients up to the age of 8 years than those aged up to 12 years and there is a significant effect on final outcome (Dadeya *et al.* 2009).

Side effects must be considered before prescribing this treatment. These effects include mental disturbances, bleeding and bruising, vivid dreams, uncontrolled movements, palpitations, fatigue and dizziness.

Pleoptics

Pleoptics is a method of treatment that is now rarely used. It is a uniocular form of treatment using after-images and Haidinger's brushes to encourage foveal fixation with normal projection. Treatment sessions need to be carried out several times per day over a period of weeks and therefore usually entails hospital admittance of the patient. Treatment requires high motivation on the part of the patient. It is used almost exclusively to treat adult patients with untreated functional amblyopia who have lost the use of the good eye (Bangerter 1969).

Of note, improvement in acuity of the amblyopic eye is quite often noted after the loss of the better eye (Vereecken & Brabant 1984). Improvement in adult amblyopic vision after loss of vision in the non-amblyopic eye may also be achieved through perceptual learning in which training incorporates grating acuity, contrast sensitivity, spatial localisation and eye-hand coordination exercises (Fronius *et al.* 2006). Furthermore, there appears to be a therapeutic effect of perceptual learning for younger patients with amblyopia regardless of whether they have received occlusion therapy or not (Liu *et al.* 2011).

Eccentric fixation

This is a uniocular condition in which there is fixation of an object by a retinal point other than the fovea without change in the principal visual direction. Eccentric fixation is seen in long-standing esotropia and microtropia with identity. The lower the level of visual acuity, the more eccentric and unstable the fixation and this is independent of size and type of stimulus (Siepmann *et al.* 2006). In cases of microtropia, the eccentric fixation should not be disrupted. However, eccentric fixation associated with large-angle deviations may be treated with occlusion. In cases of non-absolute fixation, direct occlusion is used to treat amblyopia.

With fixed eccentric fixation, indirect occlusion is commenced which involves occluding the amblyopic eye initially. Occluding the amblyopic eye disrupts fixation in the eye. Direct occlusion is then implemented full time to take advantage of the disrupted fixation and promote foveal stimulation. Red filter occlusion directly stimulates the foveal cells and may be combined with indirect occlusion (Cowle *et al.* 1967, Malik *et al.* 1969).

Cerebral visual impairment

Cerebral or cortical visual impairment involves damage to the grey and white matter of the brain. Multiple problems ensue including impaired recognition of people, objects and shapes, difficulties with orientation and spatial awareness,

difficulties with interpreting complex visual scenes and inaccurate reaching and grasping (Dutton *et al.* 1996, Rahi & Cable 2003). The condition is the commonest cause of visual impairment in children in developed countries. It is typically caused by periventricular leukomalacia, hypoxic ischaemia, hydrocephalus, meningitis, encephalitis, traumatic brain injury, metabolic disease and secondary effects of drugs and radiation (Houlihan *et al.* 2000, Dutton & Jacobson 2001, Jacobson *et al.* 2002).

The clinical characteristics of cerebral visual impairment include (McKillop & Dutton 2008):

1. Functional loss of visual acuity with crowding due to dorsal stream dysfunction.
2. Reduced contrast and colour vision.
3. Visual field loss such as homonymous hemianopia or altitudinal defects.
4. Visual inattention.
5. Simultanagnosia.
6. Loss of perception of movement.
7. Impaired accommodation and vergence.
8. Ocular motility abnormalities of strabismus, nystagmus, inaccurate saccadic and smooth pursuit eye movements.
9. Amblyopia.
10. Impaired recognition of faces, objects and shapes.

There is a high incidence of reduced accommodative responses in children with cerebral visual impairment (McClelland *et al.* 2006) and those with more severe motor impairments are at greater risk of having these deficits. Preferential looking and visual evoked potential responses are below normal for children with cerebral visual impairment although there are improvements in these indices in early childhood (Lim *et al.* 2005). Cerebral visual impairment is a risk factor for development of consecutive exotropia and surgical procedures require adaptation for children with cerebral visual impairment as recession surgery has more effect per millimetre of surgery than in normal children (van Rijn *et al.* 2009).

Delayed visual maturation

With delayed visual maturation, there is apparent blindness until approximately 12 weeks of age (Hoyt 2004). Clinical assessment of visual behaviour is classified as abnormal and infants do not fix and follow lights or targets and may have poor pupil responses. Some children display transient nystagmus that regresses over the ensuing months and disappears (Bianchi *et al.* 1998, Good *et al.* 2003). Normal acuity thresholds are shown on electrophysiological measurements of grating and vernier acuity using sweep visual evoked potentials (Good & Hou 2004). Most infants are otherwise normal, although there are cases with associated neurodevelopment delay and with ocular findings including refractive error (Winges *et al.* 2005).

The condition is proposed to be due to sub-cortical delay with improvement in vision occurring with the emergence of cortical function and cortically mediated responses (Cocker *et al.* 1998). There is thought to be due to a delay in the myelination of the visual fibres. Children usually demonstrate normal visual development thereafter.

PHACE syndrome

This syndrome consists of a large haemangioma in the face or neck and other abnormalities (Kronenberg *et al.* 2005). PHACE represents:

P – posterior fossa brain abnormalities

H – haemangioma

A – arterial blood vessel abnormalities in the head and neck

C – cardiac abnormalities

E – eye abnormalities.

The ocular abnormalities may include anterior and posterior segment abnormalities, retinal vascular abnormalities, iris vessel hypertrophy, optic nerve hypoplasia, morning glory disc appearance, microphthalmos, coloboma, congenital cataracts, exophthalmos, strabismus, amblyopia, refractive error and Horner's syndrome (Ceisler & Blei 2003, Kronenberg *et al.* 2005, Schwartz *et al.* 2006).

Size of haemangioma greater than 1 cm on the orbit or lids is considered an important predictor for development of amblyopia (Schwartz *et al.* 2006). Treatment to reduce the size of the haemangioma results in improved vision and reduction in refractive error (Schwartz *et al.* 2007).

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9

Aphakia

Aphakia follows the treatment of congenital or acquired cataract and may occur at any age, although patients are most likely to be rendered pseudophakic with the insertion of an intraocular lens following cataract extraction.

Methods of correction

Unilateral

- Intraocular lens implant
- Contact lenses
- Epikeratophakia
- Laser surgery

Bilateral

- Intraocular lens implant
- Spectacles – must be correctly centred and accurately fitted
- Contact lenses
- Laser surgery

Investigation

Case history Ascertain the cause and duration of the cataract, time of extraction and form of subsequent correction. With paediatric cases, check the time length of presence of cataract before treatment as this can help predict prognosis for visual outcome. Symptoms may include diplopia and blurred near vision. There may be a previous history of strabismus and positive family history.

An additional +3.0 DS lens in front of the corrected aphakic eye should be used for near testing with older children and adults.

<i>Visual acuity</i>	Children will generally have severe amblyopia of stimulus deprivation type and often strabismic type in addition. However, good visual acuity can be maintained in some cataracts, for example anterior peripheral lens opacity.
<i>Cover test</i>	Adults may have latent, intermittent or manifest strabismus. Children frequently have manifest secondary strabismus (see Chapter 6).
<i>Ocular motility</i>	In cases of traumatic cataract, ocular motility may be limited where there has been associated globe and/or extraocular muscle damage.
<i>Binocular function</i>	Adults may demonstrate binocular single vision with latent or intermittent strabismus. If manifest strabismus is present, the angle is corrected to assess the state of binocularity. Children rarely demonstrate binocularity due to the presence of strabismus and dense amblyopia.
<i>Aniseikonia</i>	There may be loss of fusion in adults with cataract that has been left untreated over a period of years (Sloper & Collins 1995). The presence and extent of aniseikonia can be assessed with Ruben's slides on the synoptophore or with other available methods.
<i>Angle of deviation</i>	This is measured using corneal reflections where the visual acuity is insufficient to obtain adequate fixation.

Problems with unilateral aphakia

- Insuperable aniseikonia
- Manifest strabismus:
 - Secondary divergent deviation in adults
 - Secondary convergent deviation in young children
- Stimulus deprivation amblyopia plus strabismus amblyopia

Management

Aphakia is corrected with an intraocular lens implant, contact lenses or epikeratophakia. Intraocular lenses give the least increase in image size. Spectacles may be used with bilateral cases, but problems arise with aberrations, prismatic effects, centration and weight, and they must be a good fit to encourage tolerance with young children. Contact lenses provide a wider field of view than spectacles but have a risk of infection and lack of oxygen supply to the cornea with prolonged wear.

Adults

Symptomatic heterophoria is treated with orthoptic exercises (see Chapter 3). With intermittent deviations, orthoptic exercises are used to improve fusional reserves.

Surgery may be required if a large angle is present and orthoptic exercises given post-operatively to consolidate the result. The aim is for parallel visual axes with functional cases. In cosmetic divergent deviations, aim for a slight overcorrection. Where there has been a loss of fusion, occlusion, Bangerter filters or occlusive contact lenses may be required (McIntyre & Fells 1996).

For adults with long-standing monocular aphakia and secondary strabismus, there is a significant risk of diplopia with late secondary intraocular lens implantation. Evaluation of pre-operative diplopia is essential as an inability to correct diplopia prior to strabismus surgery is predictive of persistent post-operative diplopia (Khan 2008).

Children

There is often a poor prognosis for vision in unilateral cataracts. A high incidence of ophthalmic complications, delay in establishing daily contact lens wear and a failure to achieve good compliance with occlusion therapy are factors associated with a poor visual outcome (Simons *et al.* 1999, Dewsbury 2005). However, outcomes continue to improve with earlier surgery, better optical correction of aphakia and occlusion methods (Ruth & Lambert 2006) and it is possible to achieve binocular function with early surgery and less occlusion in those with good compliance with contact lens and occlusion therapy (Brown *et al.* 1999).

Contact lenses are fitted and occlusion is implemented to improve vision. These patients usually require full-time total occlusion. Where traditional occlusion has failed, occlusive contact lenses may be trialled (Joslin *et al.* 2002). Advances in contact lens technology have led to higher success rates with contact lens wear and thus improved visual outcomes (Lindsay & Chi 2010).

In cases of bilateral uneven cataracts with amblyopia, both cataracts can be removed and one intraocular lens implanted to the amblyopic eye with the better eye left temporarily aphakic as an alternative to occlusion. The secondary intraocular lens is implanted once the previous amblyopic eye has reached its potential (Yu & Dahan 2009).

Strabismus surgery may be indicated for manifest or decompensated strabismus.

The cataract can be removed within days of birth and intraocular lens (Spierer *et al.* 1999) are implanted at increasingly earlier ages with a top-up correction obtained with a contact lens.

Posterior chamber intraocular lens implantation is a safe and effective method for treatment of cataracts in children over the age of 2 years. However, there is considerable debate as to the safety and long-term effects of intraocular lens implantation in neonates.

Early treatment is advisable as this results in fewer acuity deficits than later treatment. Treatment during the initial 6 weeks is maximally effective and effectiveness rapidly decreases from 12 weeks of age (Birch *et al.* 1998). Better results are also achieved with combined treatment (intraocular lens plus contact lens) rather than sole treatment regimes.

Comparative trials of contact lenses versus intraocular lens implant for optical correction of unilateral aphakia during infancy (IATS group 2010) has shown no significant difference in visual acuity outcome between groups at age 1 year. However, additional intraocular lens operations are more frequent in the intraocular lens group. Thus, caution should be exercised when considering surgery in children younger than 6 months due to this higher incidence of adverse events (IATS group 2010).

For prognosis, the best-corrected visual acuity reaches a plateau at about 7 years of age and this is generally predictive of long-term visual outcome (Sjöstrand *et al.* 2011).

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SECTION III

10 Incomitant Strabismus

Incomitant strabismus is strabismus in which the angle of deviation differs depending upon the direction of gaze or according to which eye is fixing and is associated with defective movement of the eye or with asymmetrical accommodative effort.

Ophthalmoplegia is a group of conditions that have a variety of causative factors, where there is a paresis of two or more of the extraocular muscles.

When examining these patients, the angle of deviation may be different depending on which eye is fixing and how long the deviation has been present.

- *Primary angle of deviation:* The deviation when fixing with the unaffected eye in paralytic incomitant deviation.
- *Secondary angle of deviation:* The deviation when fixing with the affected eye in paralytic incomitant deviation.

Generally, with more recently acquired palsies, the strabismus will be quite incomitant with a significant difference between the primary and secondary angles of deviation. With time, the strabismus becomes less incomitant and it is harder to assess whether it is congenital or long standing. It is often difficult to differentiate between primary and secondary deviations, as relative concomitance usually occurs following long-standing incomitance.

Aetiology

Incomitant strabismus may be found in association with many disorders. The list below is designed to give an overall idea of the types of condition that can cause incomitance.

Congenital

Neurogenic

- In association with other congenital developmental disorders
- As an isolated feature

Form

- Aplasia; hypoplasia of nerve/nucleus
- Abnormal innervation
- Trauma during gestation/delivery
- *Inflammation:* Neonatal/antenatal

Mechanical

- Duane's retraction syndrome
- Brown's syndrome

Myogenic

- *Developmental abnormality:* Hypoplasia/hyperplasia/aplasia of extraocular muscles
- *Abnormal attachments:* For example, muscle insertions to the eyeball or to the orbital contents
- Fibrosis
- *Adhesions:* Intermuscular, muscle to orbit

Acquired

Neurogenic

- Trauma
- Inflammation, for example multiple sclerosis
- Vascular, for example hypertension
- Space-occupying lesions
- Metabolic disorder, for example diabetes

Mechanical

- *Trauma:* Development of fibrous tethers, fractures
- Space-occupying lesions
- Iatrogenic adhesions
- Secondary to myogenic inflammation, for example thyroid eye disease

Myogenic

Lesions at:

- neuromuscular junction, for example myasthenia gravis
- muscle fibre membrane, for example myotonia
- muscle fibre contents, for example dystrophies (chronic progressive external ophthalmoplegia), endocrine (dysthyroid eye disease), inflammatory (myositis)

Table 10.1 Differences between congenital and acquired defects.

	Congenital	Acquired
Presentation	Symptoms of decompensation Unaware of abnormal head posture Unacceptable cosmetic appearance	Diplopia and occasionally pain Aware of uncomfortable abnormal head posture
Ocular motility	Often full muscle sequelae	Muscle sequelae not fully developed
Binocular function	Extended vertical fusion range	Normal fusion range

Aid to diagnosis

Congenital and acquired defects often show different characteristics, which help in making the diagnosis. Table 10.1 shows the findings that might be expected when the patient is examined in the eye clinic.

There are also differences between neurogenic and mechanical defects, which are summarised in Table 10.2.

Table 10.2 Differences between neurogenic and mechanical defects.

	Neurogenic	Mechanical
Cover test	Deviation in primary position reflects the extent of palsy	Often only small deviation in primary position
Ocular motility	Movement is greater on ductions compared to versions No retraction of the globe	Movement is the same on ductions and versions Retraction may be noted
Hess chart	Space between inner and outer fields is equal and proportional Fields are displaced away from position of greatest limitation	Outer field is displaced close to inner field in the position of greatest limited movement
Forced duction test	Full passive movement	Limited passive movement
Intraocular pressure	Same in all positions of gaze	Increases when looking away from the position of limitation

Diplopia

Diplopia may be binocular or monocular and may be horizontal, vertical and/or torsional.

Causes of diplopia

Refractive error	Uncorrected Aniseikonia
Strabismus	Concomitant secondary strabismus Concomitant strabismus with loss of suppression Acquired strabismus due to cranial nerve palsy, trauma, systemic disease, iatrogenic (Trobe 1984)
Fusion deficit	Decompensation of long-standing heterophoria, acquired central fusion loss (Pratt-Johnson & Tillson 1988)
Retinal pathology	Retinal folds, retinal membrane, retinal traction (dePoole <i>et al.</i> 2005)
Visual field loss	Impairment of retinal correspondence with severe visual field loss, hemifield slide phenomenon (Rowe 1996)

Investigation

<i>Case history</i>	Question associated visual symptoms. Metamorphopsia, blur, distortion and illusory movements are typical of retinal pathology. History of closed head injury may indicate cranial nerve palsy or acquired central fusion disruption.
<i>Diplopia type</i>	Ascertain whether horizontal crossed (typical of exo-deviations) or uncrossed (typical of eso-deviations). Where diplopia is opposite to that expected, consider the presence of paradoxical diplopia due to sensory changes in long-standing concomitant strabismus (Castleberry & Arnoldi 2003). Torsional diplopia is typically seen with fourth nerve palsy (Woo <i>et al.</i> 2005). Impact of diplopia may be ascertained by questionnaire (Holmes <i>et al.</i> 2005).
<i>Occlusion</i>	Retinal diplopia will remain under monocular conditions whereas binocular diplopia will be eliminated with occlusion of one eye (Records 1980).
<i>Pinhole test</i>	Monocular diplopia is often relieved when looking through a pinhole.
<i>Area of diplopia</i>	The area of diplopia (and field of binocular single vision) can be assessed by plotting single versus diplopic vision using the Goldmann perimeter, arc perimeter or cervical range of motion test (Holmes <i>et al.</i> 2005, Rowe & Hanif 2011).

Management

Diplopia may be joined with Fresnel prisms, which may later be incorporated into a glasses prescription. For larger angles of deviation, use of botulinum toxin

or strabismus surgery (relevant to the condition) may join the diplopic images. Occlusion can be used in total or partial form to eliminate diplopic images. For long-term management, a small number of patients opt for an occlusive contact lens or occlusive intraocular lens implant (Kwok & Watts 2009). Intraocular occlusive lenses are also now designed to allow high-level transmission such that equipment such as scanning laser ophthalmoscope and optical coherence tomography can be used to image the posterior segment of the eye (Yusuf *et al.* 2011). A further, unusual alternative is that of a corneal tattoo to block diplopia (Dawson *et al.* 2009).

Abnormal head posture

An abnormal head posture is any state in which the head is consistently not held in an upright position but adopts one or more of the following components (Fitton 1951, Nutt 1963):

- Head tilt
- Face turn
- Chin elevation or depression

Head tilt

The tilt is generally towards the hypotropic eye and may be adopted to utilise the torsional movements of the eyes.

Face turn

This is adopted to place the eyes away from field of action of the paresed muscle for vertically acting muscles as well as horizontal. It also enables the eyes to fix centrally where there is gross limitation of movement.

Chin elevation or depression

This may be adopted to place eyes away from the field of action of the affected muscle or to utilise other muscles having the same action. It can also be used for comfort in some mechanical limitations where movement is painful.

Assessment of head posture

The following factors should be considered:

1. Ensure sitting upright or standing.
2. Observe head posture from directly in front of the patient.
3. Note facial symmetry/asymmetry.
4. Straighten head and allow patient to assume usual position.
5. Assess near and distance.
6. May be intermittent; therefore, observe throughout.
7. Examine old photographs in suspected long-standing cases.

8. Scoliosis of spine seen in congenital or early onset tilt.
9. Compare which ear is more visible.
10. Check whether eyes are level.
11. Observe for chin position from side if not obvious.

Uses of the Abnormal Head Posture

Diagnostic

1. Diagnosis of the affected muscle in palsies.
2. Differential diagnosis of congenital and acquired palsies by the following:
 - a. Awareness of presence of a head posture
 - b. Degree of head posture in relation to amount of limitation of movement
 - c. Examination of old photographs
3. Presence of normal binocular functions in paralytic squint. An abnormal head posture without binocular single vision may indicate pre-existing binocular single vision.
4. Use of the Bielschowsky head tilt test to differentiate the affected muscle.

Therapeutic

A marked abnormal head posture may be an indication for surgery in the following:

1. Ocular palsies
2. Musculofacial anomalies
3. Ptosis
4. Nystagmus

Reasons for abnormal head postures

Non-ocular

1. Non-ocular torticollis (contracture of the sternomastoid muscle)
2. Deafness
3. Arthritic and rheumatoid conditions
4. Habit
5. Shyness
6. Mental development delay
7. Presbyopia
8. Disorders of the cervical spine

Ocular (Wesson 1964)

1. To enable the development of binocular single vision in cases of congenital paralytic strabismus and musculofacial anomalies.

2. To maintain binocular single vision in cases of acquired paralytic strabismus, mechanical limitations and A or V patterns.
3. To overcome symptoms other than diplopia such as avoiding painful ocular movements or in cases of physiological V pattern.
4. To gain foveal fixation in cases of marked infantile esotropia, bilateral lateral recti paralysis and in myogenic conditions with grossly limited movement.
5. To improve visual acuity in cases of bilateral ptosis, nystagmus, under- or overcorrected spherical refractive errors, wrongly corrected astigmatism and visual field defects.
6. To separate diplopia in paralytic strabismus by moving the images further apart so that the second image can be ignored.
7. In dissociated vertical deviation, there may be a tilt or turn towards the fixating eye.
8. To protect the eyes.
9. To compensate for visual field loss.

Differential diagnosis between ocular and non-ocular torticollis

Differential diagnosis can be aided by evaluation of changes in ocular deviation and eye movements with a number of assessments as outlined below in Table 10.3.

Table 10.3 Torticollis differential diagnosis.

	Ocular	Non-ocular
Aetiology	Vertical extraocular muscle palsies	Contracture of sternomastoid muscle
Effect of straightened head	Vertical deviation decreases and may decompensate	Shoulder raises
Ocular movements	Over- and underactions of vertically acting muscles in the same direction as the face turns	Full or unrelated to head posture
Ocular deviation	Vertical and may have a horizontal and torsional component	None or not related to head posture
Type of head posture	Depends on affected muscle	Tilt to affected side Turn to opposite side
Head tilt	Of varying degree	Usually marked
Effect of occlusion	Head tilt reduced or is abolished with one eye	Head tilt persists when one eye is occluded or when both eyes are open
Treatment	Surgery on the vertically acting muscles may succeed in abolishing or reducing the head posture	Treatment of the neck defects Physiotherapy

Conditions in which abnormal head postures are present

1. *Bilateral ptosis*: To achieve binocular single vision and better visual acuity.
2. *Incomitant squint*:
 - *Neurogenic*: To achieve binocular single vision or separate diplopic images where there has been a traumatic loss of fusion. A marked face turn makes use of the nose as an occluder.
 - *Mechanical*: To avoid pain, achieve binocular single vision or separate the diplopic images.
3. *Nystagmus*: To make use of a null point. Face turn adopted.
4. *Physiological V*: Alteration in chin position.
5. A and V patterns.
6. *Heavy eye syndrome*: Head tilt.
7. *Heterophoria*: A large heterophoria in the primary position may decrease on elevation or depression.
8. *Manifest deviation*: A small angle deviation may be controlled on elevation or depression.
9. *Tripartite field*: Abnormal head posture to cross-fixate.
10. *Monocular blindness, occlusion, visual field defects*: Abnormal head posture to centralise the field of vision.
11. *Uncorrected refractive errors*: Abnormal head posture to achieve better vision.
12. *Ill fitting spectacles/incorrect cylinder*: Head tilt.

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11 A and V Patterns

A and V patterns are patterns of horizontal incomitance in which a significant increase or decrease in the angle of horizontal deviation is noted between an elevated and depressed position of gaze.

Classification

- *A pattern* (Fig. 11.1): This is a condition in which there is relative convergence on upgaze and relative divergence on downgaze. There must be a minimum of 10-prism dioptres difference between upgaze and downgaze.
- *V pattern* (Fig. 11.2): This is a condition in which there is relative divergence on upgaze and relative convergence on downgaze. There must be a minimum of 15-prism dioptres difference between upgaze and downgaze. This allows for a slight physiological V pattern.

Variants of A and V patterns include:

- *X pattern*: There is relative divergence on both up- and downgaze.
- *Y pattern*: There is relative divergence on upgaze with no significant difference between the primary position and downgaze.
- *λ pattern*: There is relative divergence on downgaze with no significant difference between the primary position and upgaze.
- *\diamond pattern*: There is relative convergence on both up- and downgaze.

Aetiology

There are various theories relating to the aetiology of A and V patterns (Folk 1997). These may involve imbalance or abnormal insertions of horizontally acting extraocular muscles, imbalance of vertically acting extraocular muscles, sagittalisation or structural anomalies. Horizontal innervation muscle theories are based



Figure 11.1 'A' pattern.

on the concept of physiological V pattern in which medial recti have greater power on downgaze and lateral recti have greater power on upgaze. Many have reported anomalies in the insertions of the horizontal recti muscles; thus, if the muscle insertions are higher or lower than the normal position, abduction or adduction is subsequently increased on upgaze or downgaze. Raised insertion of the medial rectus has been found at the time of surgery in patients with elevation on adduction. Postic (1963) and Piper (1963) reported anomalies of the rectus muscle insertions particularly in patients with A and V patterns with additional anomalies of the lid fissures. Kushner (1985) reported that if muscles are displaced, they take on additional actions.

Brown (1953) reported A and V patterns caused by primary anomalies in function of vertical muscles. Urist (1958) stated A and V patterns may be caused by anomalies

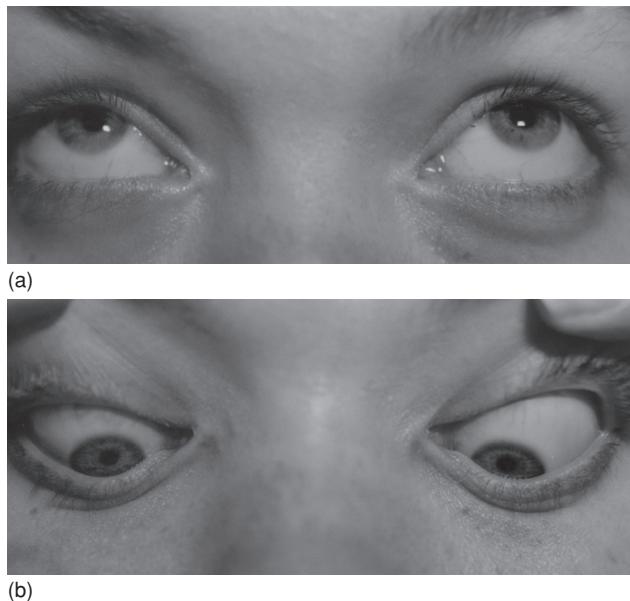


Figure 11.2 'V' pattern.

of cyclovertical muscle actions. Fink (1959) found anomalies in the muscle places of the superior oblique tendon and the inferior oblique muscle during anatomical dissection of the orbits.

Normally, the planes of both the superior and inferior obliques are identical and form an angle of 51° with the anterior–posterior axis of the globe. If one insertion is more posterior than normal, the muscle axis will lie in a more sagittal place, that is, it is closer to the anterior–posterior axis of the globe. The effect of this 'sagittalisation' is to reduce the muscle's torsional action and there is muscle overaction to compensate (Gobin 1968).

Urrets-Zavalia (1961) believed that anomalies in action of the cyclovertical muscles, which may cause variations of the horizontal deviations in up- and downgaze, are very different in patients with mongoloid and anti-mongoloid features. Mongoloid features include hyperplasia of malar bones, upwards slanting of palpebral fissures and a straight lower lid margin. Anti-mongoloid features include hypoplasia of malar bones, downwards slanting of palpebral fissures and an s-shaped contour of the lower lid margin.

Aetiology may also be related to muscle pulleys (Clark *et al.* 1998, Heo *et al.* 2008). Apparently overacting inferior obliques or superior obliques have shown difference in positions of lateral and medial rectus muscle pulleys. The heterotopia alters the vertical position of the pulleys, thus changing the course and the actions of the lateral rectus and medial rectus muscles, which, in turn, produces elevation or depression in adduction of the fellow eye depending on whether the pulley is displaced inferiorly or superiorly. Abnormal actions of vertical recti muscles are also reported (Ozkan *et al.* 2003).

A eso pattern

1. Lateral rectus underaction results in less abduction on elevation.
2. Low medial rectus insertions result in more adduction on elevation due to different muscle position and increased muscle tension.
3. Inferior oblique underaction results in less abduction on elevation. The superior oblique will overact resulting in more abduction on depression.
4. An anterior inferior oblique muscle insertion will result in less abduction on elevation.
5. Superior oblique sagittalisation results in superior oblique overaction.
6. Mongoloid facial features (Hatsukawa *et al.* 2001).
7. High tone of superior oblique muscles present with long-standing eso-deviations results in superior oblique overaction.

A exo pattern

1. Medial rectus underaction results in less adduction on depression.
2. High lateral rectus insertions result in more abduction on depression.
3. Inferior rectus underaction results in less adduction on depression. The superior rectus will overact resulting in more adduction on elevation.
4. Superior oblique sagittalisation results in superior oblique overaction.
5. Anti-mongoloid facial features.
6. High tone of superior rectus muscles present with long-standing exo-deviations results in superior rectus overaction.

V eso pattern

1. Medial rectus overaction results in more adduction on depression.
2. High medial rectus insertions result in more adduction on depression.
3. Superior oblique underaction results in less abduction on depression. The inferior oblique will overact resulting in more abduction on elevation.
4. Inferior oblique sagittalisation results in inferior oblique overaction.
5. An anterior superior oblique muscle insertion will result in less abduction in depression.
6. Anti-mongoloid facial features.
7. High tone of inferior oblique muscles present with long-standing eso-deviations results in inferior oblique overaction.

V exo pattern

1. Lateral rectus overaction results in more abduction on elevation.
2. Low lateral rectus insertions result in more abduction on elevation.
3. Superior rectus underaction results in less adduction on elevation. The inferior rectus will overact resulting in more adduction on depression.
4. Inferior oblique sagittalisation results in inferior oblique overaction.

5. Mongoloid facial features.
6. High tone of inferior rectus muscles present with long-standing exo-deviations results in inferior rectus overaction.

Investigation

Aims

1. To detect and measure A/V patterns.
2. To assess ocular movements associated with A/V patterns.
3. To assess significance of A/V patterns for prognosis and management.

Criteria for diagnosis

This is based on the amount of difference in measurement of the horizontal angle of deviation:

1. V pattern; minimum difference of 15 prism dioptres from upgaze to downgaze
2. A pattern; minimum difference of 10 prism dioptres from upgaze to downgaze (Knapp 1959).

There is a physiological tendency to relatively diverge in upgaze, and thus the minimum standards required for a V pattern is larger than that for an A pattern.

Significance of A/V patterns

1. *Diagnosis of incomitance*: The pattern may be the first sign of incomitance and not just a horizontal squint.
2. *Diagnosis of paretic element*: Strabismus may not have been noticed but A/V pattern indicates the presence of early onset squint.
3. Indication of possible vertical element in apparent horizontal deviation.
4. Suggestion of bilateral ocular muscle imbalance.
5. Binocular single vision may be present in the position of minimal deviation even if it is not present in the primary position. Therefore, test in that position.
6. Cosmesis is more significant in certain positions such as primary gaze and elevation but less so for downgaze as the lids cover the eye position.

Incidence/prevalence

There is a varied incidence of A and V patterns reported in the literature. However, it is accepted that the incidence is around 20%. The relative frequency of various types of patterns is not clearly established but thought to be, in order of greatest to least frequent, V eso, A eso, V exo, A exo.

A and V patterns are especially prevalent in certain conditions such as;

1. *Infantile esotropia*: High incidence of both A and V patterns along with dissociated vertical deviation (DVD) and inferior oblique overactions.
2. *Duane's retraction syndrome*: V pattern with upshoots and A pattern with downshoots.
3. *Brown's syndrome*: Most have V pattern explained by increased abduction resulting when mechanically inhibited inferior oblique receives increased innervation to elevate the eye.
4. *Acquired fourth nerve palsy (particularly bilateral)*: V pattern esotropia.
5. *Thyroid eye disease*: A pattern exotropia may result from over recession of the inferior recti muscles or from orbital decompression.

<i>Case history</i>	An abnormal head posture may be noted. Strabismus and abnormal eye movements may have been observed and patients may present with symptoms of decompensation.
<i>Abnormal head posture</i>	Chin elevation is seen with A eso and V exo patterns and chin depression with A exo and V eso patterns. In addition, a face turn and head tilt may be present due to associated extraocular muscle paresis.
<i>Cover test</i>	<p>This should be performed with and without the abnormal head posture to assess the state of compensation or decompensation of the deviation.</p> <p>Perform in primary position, upgaze and downgaze for both near and distance.</p> <p>Control accommodation by using an accommodative target for fixation and ensuring it is seen clearly, and by testing with glasses worn, particularly if hypermetropic. Repeat without glasses as frames may restrict fixation on up and downgaze.</p> <p>Failure to wear appropriate correction or use an accommodative target may result in true patterns being concealed (Knapp 1959, von Noorden & Olsn 1965).</p>
<i>Ocular motility</i>	A and V patterns are common in some motility syndromes as detailed above. One should check for asymmetry of bilateralism in ocular movements. Where overacting extraocular muscles are noted and the corresponding underacting muscles should be assessed.
<i>Binocular function</i>	<p>If the patient is hypermetropic, use an accommodative target to assess in up and downgaze. DVD may also be noted.</p> <p>A good level of binocular function may be maintained with an abnormal head posture. If negative in the primary position, test in the position of minimal deviation.</p> <p>If no free space binocularly can be demonstrated, test on the synoptophore. If negative in the primary position, move tubes into the position of minimum deviation.</p>

Angle of deviation

Measurements should be made in up and downgaze to confirm presence and assess if it is diagnostically significant.

There must be a difference of 15 prism dioptres between measurements on elevation and depression in cases of V patterns. This allows for a physiological V pattern. There must be a difference of 10 prism dioptres in cases of A patterns to confirm the diagnosis. Accommodation must be controlled when measuring the nine positions of gaze as a pseudo A or V pattern may result or a pattern can be masked if accommodation is exerted or not (Knapp 1959, von Noorden & Olsen 1965). Therefore, measurements are preferable at distance fixation to prevent the influence of accommodation.

Measurements may be made using the synoptophore and moving the tubes or with the prism cover test by moving the head or the target. The synoptophore is a repeatable test but does not always reveal the full difference in the angle of deviation.

Knapp questioned whether errors in measurement were incurred by measuring the deviation with the head tilted backwards and forwards to obtain depression and elevation of the eyes or with the head fixed while the patient fixates above and below the horizontal plane. However, there has generally been little difference found whether the target or head is moved (Magee 1960, Scott 1968).

Breinin (1961) stated that the eyes should not be rotated in extreme positions since the mechanical effects of the check ligaments and musculo-facial system may alter the deviation.

Confirm A/V patterns and associated over- and underactions.

A and V patterns are revealed by sloping sides to the two fields.

Hess chart and field of binocular single vision

Management

It is important to determine whether the strabismus is functional, to consider the cosmetic aspect or if functional, to differentiate between compensated and decompensated and the extent of the head posture. Give the refractive correction as appropriate.

Functional cases

Management consists of monitoring the patient to ensure that comfortable symptom free binocular single vision is maintained without an excessive head posture. If an abnormal head posture is present of a significant degree, surgery must be considered to eliminate this. Surgery may also be undertaken for upshots. It is important to ensure there are no factors that may contribute to decompensation such as a

reduced near point of convergence. Prisms are indicated where there are signs of decompensation with intermittent diplopia and loss of control. Prisms may be used diagnostically, provided the incomitance is not excessive.

Corrective cases

Surgery is considered only for large deviations, marked abnormal head postures and upshoots. An increase in angle of deviation on depression is masked by the upper eyelids. Obvious strabismus on elevation may be cosmetically poor in children but is likely to be less obvious in adult life. In such cases, the height of family members should be considered.

Aims of surgery

1. Correct horizontal angle in primary position.
2. Reduce incomitance in vertical gaze.
3. Eliminate any noticeable upshoot on lateroversion.

General principles

1. Bilateral symmetrical procedures.
2. May require more than one operation.
3. If no vertical extraocular muscle abnormality is present, operate on horizontal recti. If vertical extraocular muscle abnormalities are documented, then surgery on the vertical muscles should be considered.

Surgery on horizontal recti

This is recommended when there is no evidence of vertical extraocular muscle dysfunction or minor vertical muscle incomitance is present only in extreme positions of elevation or depression.

- *Transpositions:* Vertical transposition of horizontal rectus muscles may be combined with a recession and less commonly with a resection. This is based on the principle that with the eyes in elevation or depression, the muscle plane, determined by the centre of rotation of the globe and centres of origin and insertion, changes. Extraocular muscles are usually displaced at a distance, which is equivalent to a muscle width or half a muscle width.
- *Boyd's technique:* Recessions are further enhanced by recessing the upper or lower margin of the horizontal rectus more than the opposite margin (Ohba *et al.* 2004).

Surgery on vertically acting muscles

- Weakening procedures; recession or myectomy, tenotomy or tenectomy (Urist 1951, Villaseca 1961)

- Strengthening procedures; plication (tuck) or resection or Harado-Ito
- Anteropositioning
- Transplant vertical recti; nasal or temporal muscle displacement (Miller 1960, von Noorden 1963)

A eso pattern

- Bilateral medial rectus recessions, moving the insertions up (Goldstein 1967, Urist 1968, Garrido *et al.* 2004).
- Sloping medial rectus recessions with maximum recession on the upper border of muscles (Boyd *et al.* 1971).
- Weaken the superior oblique by tenotomy or tenectomy (Shin *et al.* 1996, Lee & Rosenbaum 2003, Ron *et al.* 2009).
- Superior oblique anteropositioning.
- Transpositioning of the superior rectus temporally (Miller 1960).

A exo pattern

- Bilateral lateral rectus recessions, moving the insertions down (Urist 1968, Scott *et al.* 1989).
- Sloping lateral rectus recessions with maximum recession on the lower border of muscles (Boyd *et al.* 1971).
- Weaken the superior oblique by tenotomy or tenectomy (Shuey *et al.* 1992, Ron *et al.* 2009). Posterior tenotomy of the superior oblique weakens the vertical action of the muscle without significantly affecting the torsional action (Sharma *et al.* 2005).
- Superior oblique anteropositioning (Caldeira 1975, Buckley & Flynn 1983, Romano & Roholt 1983, Souza-Dias & Uesugui 1986).
- Transpositioning of the inferior rectus medially.

V eso pattern

- Bilateral medial rectus recessions, moving the insertions down (Urist 1951, Goldstein 1967, Oya *et al.* 2009).
- Sloping medial rectus recessions with maximum recession on the lower border of muscles (Boyd *et al.* 1971).

Both slanting recessions and horizontal transposition surgery have been found to correct V eso patterns with no significant difference between the procedures (Mostafa & Kassem 2010).

- Weaken the inferior oblique (Billet & Freedman 1969) by anterior transposition, graded recession or hang-back recession (Minguini *et al.* 2004, Kumar *et al.* 2008).
- Strengthen the superior oblique with a tuck procedure (Hiles *et al.* 1985).
- Inferior oblique anteropositioning (Gobin 1964).
- Tranpositioning of the inferior rectus temporally (Miller 1960, von Noorden 1963).

V exo pattern

- Bilateral lateral rectus recessions, moving the insertions up (Urist 1968, Oya *et al.* 2009).
- Sloping lateral rectus recessions with maximum recession on the upper border of muscles (Boyd *et al.* 1971).
- Weaken the inferior oblique by anterior transposition or graded recession (Minguini *et al.* 2004).
- Inferior oblique anteropositioning.
- Transpositioning of the superior rectus medially (Miller 1960).

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12 Accommodation and Convergence Disorders

Accommodation is the ability to increase the convexity of the intraocular lens in order to obtain a clear image of a near object. It is associated with convergence and pupil constriction. Circular muscles of the ciliary body contract and this brings about accommodation. The ciliary body is supplied by the III nerve and parasympathetic nerve supply. Brücke's muscles are meridional muscles that contract when looking into the distance and are supplied by sympathetic nerves.

Accommodative disorders

A defect of the focusing mechanism may result in:

- deficient accommodation
 - presbyopia (physiological, premature)
 - accommodative weakness (insufficiency, fatigue)
 - accommodative paralysis
- excessive accommodation
 - accommodative spasm
- failure to alter accommodation
 - accommodative inertia.

Presbyopia – physiological

Aetiology

Normal ageing changes of the crystalline lens.

Investigation

Refraction A near addition is often required.

Case history Blurred vision is experienced for reading print especially in poor light. Asthenopic symptoms are common. Symptoms depend on age,

	refractive error (earlier in hypermetropia), the individual's normal reading position and requirements for close work.
<i>Cover test</i>	Often exophoric. This increases in angle when wearing the reading correction and may decompensate or become an intermittent near exotropia. Patients may have an altered fixation pattern (Oystreck & Lyons 2003). During early presbyopia, patients may have an esotropia due to increased convergence tonus linked to increased accommodative effort (Wright <i>et al.</i> 2005).
<i>Binocular function</i>	The control of the deviation must be assessed at near fixation. It is important to assess the base-out fusion range for exo-deviations and base-in fusion range for eso-deviations to assess control.
<i>Convergence</i>	There may be a reduced near point.
<i>Accommodation</i>	This should be checked against the expected age-matched result.

Management

The minimum convex lenses are given to rid symptoms taking into account the reading and near working position. Prisms may be added to the spectacle correction if there is an associated uncompensated heterophoria. However, if the angle of deviation is large, this may warrant treatment with botulinum toxin or surgery (see Chapter 5).

Presbyopia – premature (non-physiological)

Aetiology

Lens changes, glaucoma, poor general health and drugs. This may be permanent or temporary depending on the cause.

Investigation

<i>Refraction</i>	A near addition is often required.
<i>Case history</i>	Blurred vision is noted for reading print with asthenopic symptoms.
<i>Visual acuity</i>	Check for improvement in visual acuity with a pinhole test.
<i>Cover test</i>	Often exophoric. This increases in angle when wearing the reading correction and may decompensate or become an intermittent near exotropia.
<i>Binocular function</i>	The control of the deviation must be assessed at near fixation. It is important to assess the base-out fusion range for exo-deviations and base-in fusion range for eso-deviations to assess control.

Management

A permanent spectacle correction is only given if the cause cannot be treated. However, it may be necessary to give a temporary correction while the cause is being treated.

Accommodative insufficiency

This is a deficiency in the ability to obtain the amount of accommodation that would be expected from the patient's refractive state and age (Henshall & Rowe 2002).

Aetiology

Drugs (high oestrogen contraceptive pill, tranquillisers, valium, anti-hypertensive drugs and atropine-based drugs), poor general health, history of prolonged fever, vitamin deficiencies, virus infection and trauma (Mazow & France 1989).

Investigation

<i>Refraction</i>	A near dynamic retinoscopy may show a difference between the accommodative demand required for the stimulus versus the actual response with a lag of accommodation noted.
<i>Case history</i>	Blurred vision is noted for near vision with general asthenopia and micropsia if there is marked insufficiency.
<i>Visual acuity</i>	Distance vision is normal with poor near vision.
<i>Cover test</i>	An exophoria is demonstrated for near fixation with good recovery becoming esophoria on extreme effort to accommodate.
<i>Convergence</i>	Over convergence is possible, which improves with convex lenses. Accommodative insufficiency may also be associated with convergence insufficiency (Shin <i>et al.</i> 2011).
<i>Accommodation</i>	Pupils should be carefully assessed to ensure normal reaction. This is assessed unioocularly and binocularly, and also retested with concave lenses. Accommodative facility (typically using ± 2 dioptre lenses) is reduced, particularly with the minus lens phase of the test. This is diagnostic of accommodative insufficiency (Cacho <i>et al.</i> 2002). The condition normally affects both eyes but if one eye is affected, a local cause should be suspected, such as: <ul style="list-style-type: none"> • cyclitis • glaucoma, lens change • trauma.

Management

Elicit the cause and treat this where possible. The refractive error is corrected giving the minimum convex lenses on a temporary or permanent basis. The plus lens

addition reduces the blur to an extent that the remaining blur can be compensated by the patient's accommodative capacity and encourages the accommodative system to regain normal capacity. A +1 DS addition is shown to be effective whereas a +2 DS addition reduces the blur too much to encourage the patient's accommodative capacity to compensate (Wahlberg *et al.* 2010).

Miotics may be of use to increase the depth of focus. By reducing the pupil size, less accommodative effort is required, which helps to alleviate symptoms. Orthoptic exercises may be of benefit with mild insufficiency and where associated with convergence insufficiency. Treatment with flipper lenses aids improvement of accommodative amplitude (Brautaset *et al.* 2008).

Accommodative fatigue

There is a deficiency in the ability to maintain the amount of accommodation that would be expected from the patient's refractive state and age. Accommodation is sufficient but tires easily and therefore differs from accommodation insufficiency, which is a more constant condition.

Aetiology

Prolonged close work, poor general health, general fatigue, drugs, hysterical reaction and uncorrected refractive error.

Investigation

<i>Visual acuity</i>	Distance vision is normal but near vision deteriorates with time.
<i>Cover test</i>	Esophoria for near fixation is demonstrated.
<i>Convergence</i>	Poor.
<i>Accommodation</i>	The range is poor for the patient's age, deteriorating on repeated testing. A poor response is noted with flipper lenses.
<i>Binocular function</i>	A poor fusion range is demonstrable particularly the convergent range.

Management

This is a temporary condition that improves with rest and restoration of good general health. The refractive error should be corrected. Temporary convex lenses may be used to alleviate symptoms. Orthoptic exercises to improve the fusional range may be necessary.

Accommodative paralysis

There is a total inability to accommodate due to ciliary paralysis.

Aetiology

Drugs (anticholinergic cycloplegic), toxins (botulism), trauma (direct injury to the eye, head trauma and whiplash), congenital (e.g. III nerve palsy) and neurogenic (III nerve palsy, late feature of diphtheria, progressive midbrain disorder and pineal tumour).

Investigation

<i>Case history</i>	The severity of the symptoms depends on the refractive error. An emmetrope will see clearly at distance fixation but not for near fixation. Hypermetropes see little at any distance and myopes have mild symptoms.
<i>Visual acuity</i>	Near vision is defective.
<i>Cover test</i>	Exotropia is seen at near fixation or exophoria depending on the state of convergence.
<i>Convergence</i>	There may be an associated convergence paralysis.
<i>Accommodation</i>	Complete loss of accommodation with fixed dilated pupil is demonstrable. It usually affects both eyes and, if uniocular, most commonly indicates a III nerve palsy.
<i>Binocular function</i>	Reduced binocularity is noted at near fixation particularly stereopsis.

Management

Treat the cause if possible. The refractive error is corrected and convex lenses and base-in prisms are given for close work. The minimal strength prism should be given. Miotics are useful for a partial palsy to increase the depth of focus.

Accommodative spasm

A condition in which the ciliary muscle is contracted and cannot be relaxed. As a result, accommodation is continuously exerted.

Aetiology

Functional cases may have an underlying emotional cause (Rutstein *et al.* 1988), uncorrected moderate degree of hypermetropia, effort to control intermittent divergent strabismus, overwork and fatigue or temporary spasm from miotics. Organic cases are due to irritative lesions of the brain stem, trigeminal neuralgia, drugs (morphine and vitamin B), multiple sclerosis, trauma and cyclic oculomotor spasm (Dagi *et al.* 1987, Goldstein & Schneekloth 1996, Monteiro *et al.* 2003, Sitole & Jay 2007).

Investigation

<i>Case history</i>	Mild symptoms involve difficulty in relaxing accommodation leading to blurred vision (pseudo myopia). Marked symptoms include pseudomyopia, pain and tenderness of the globe, macropsia, headaches and an inability to concentrate.
<i>Refraction</i>	Most patients are emmetropic under cycloplegic examination.
<i>Visual acuity</i>	Mild cases note that vision is better without a hypermetropic correction. Marked cases note severe loss of distance vision and near vision can also be affected. Vision improves with a pinhole or concave lenses.
<i>Cover test</i>	Mild cases are usually esophoric in the distance. Marked cases have an esotropia in the distance with diplopia and spasmotic miosis.
<i>Ocular motility</i>	Jerky movements are often noted with defective abduction.
<i>Convergence</i>	There is usually an associated convergence spasm.
<i>Accommodation</i>	Excessive accommodation is detected and this is usually a bilateral condition.

Management

Treat the underlying cause where possible. Correct the refractive error but initially undercorrect hypermetropes and then gradually increase. Marked spasms are treated with atropine for several weeks and the atropine is then gradually reduced. Where atropine is not tolerated, alternative mydriatics may be tried, such as cyclopentolate. However, these are less effective and generally not well tolerated either.

It is possible to give orthoptic exercises to increase negative relative convergence once the spasm has relaxed and this may help to prevent recurrence.

Where there is an associated convergence spasm, botulinum toxin may be used. Some forms of spasm may be a type of dystonia and have been reported to respond well to botulinum toxin and also to medial rectus recession (Kaczmarek *et al.* 2009, Noonan *et al.* 2001).

Clear intraocular lens extraction has been proposed for persistent accommodative spasm and pseudomyopia (McMurray *et al.* 2004).

Accommodative inertia

This is a condition in which there is difficulty in changing the accommodative state from one fixation distance to another. Patients experience difficulty in changing focus quickly.

Aetiology

Poor general health, fatigue, unequal refractive errors and early presbyopia. If unilateral, the likely diagnosis is Holmes-Adie syndrome.

Investigation

<i>Case history</i>	The condition is more common over the age of 30 years. Intermittent blurring of vision is noted for near fixation and/or distance.
<i>Visual acuity</i>	Initially, blurred vision for near vision is noted. This eventually clears and then becomes blurred for distance due to slow relaxation.
<i>Cover test</i>	Patients may show signs of decompensation of an exophoria.
<i>Accommodation</i>	The range is below normal for the patient's age for both amplitude of accommodation and facility of accommodation.
<i>Binocular function</i>	Patients may have reduced fusion amplitude.

Management

The refractive error is corrected and general health should be improved. The fusion range and convergence may be returned to normal levels with orthoptic exercises.

Micropsia

Objects appear smaller than their natural size. If excessive accommodative effort is exerted, the brain will interpret that the retinal image is produced by an object close to the eye and therefore this will appear smaller than natural size, that is, micropsia occurring in accommodative palsy.

Macropsia

Objects appear larger than their natural size. If reduced accommodative effort is required, the brain will interpret that the object producing the retinal image is further away and, therefore this will appear larger than normal size, that is, macropsia in accommodative spasm.

Convergence anomalies

Convergence is the mechanism that keeps the visual axes directed towards the object.

A defect in the convergence mechanism may result in:

- convergence insufficiency
- convergence paralysis
- convergence spasm.

Convergence insufficiency

This is the inability to obtain and/or maintain adequate binocular convergence without undue effort. Convergence insufficiency may be a primary or secondary insufficiency (Mazow & France 1989, Rowe *et al.* 2001).

Aetiology

Primary

Pre-disposing causes include a wide interpupillary distance, occupations using mainly distance vision or occupations using mainly uniocular vision.

Precipitating causes include illness, ocular fatigue from prolonged close work and poor lighting, examinations, advancing age, drugs and pregnancy.

Secondary

Secondary or organic convergence insufficiency is reported due to a variety of factors. A number of reports associate convergence insufficiency with whiplash injuries following road traffic accidents (Anderson 1961, Greaves 1987) where the aetiology is presumed to be at brainstem level. Presence of convergence insufficiency and near exotropia can be a soft sign for neurological disease (Phillips *et al.* 2005).

Other associated factors include refractive error such as uncorrected myopia or correction of presbyopia, iatrogenic (weakened medial rectus), poor general health, neurological impairment due to multiple sclerosis or space occupying lesions, heterophoria (convergence weakness exophoria), vertical deviation, accommodative insufficiency and defective fusion (Lyle & Wybar 1970, Schwyzer 1978, Mills 1994, Wicks 1994, Rowe *et al.* 2001). Cases of convergence insufficiency have also been reported with superior oblique myokymia (Waddingham 1995).

Investigation

<i>Case history</i>	Symptoms are those of headache, difficulty in changing focus, blurring of print, intermittent diplopia, reading difficulty, asthenopia (Scheiman <i>et al.</i> 2011). The patient may have had treatment before.
<i>Visual acuity</i>	Near vision may be reduced if accommodation is involved.
<i>Cover test</i>	An exophoria is usually present on cover testing and tends to be of the convergence weakness type.
<i>Ocular motility</i>	Associated ocular motility abnormalities such as superior oblique underaction can cause decompensation and reduced convergence near point (Rowe <i>et al.</i> 2001).
<i>Convergence</i>	There is a reduced level of convergence that should be assessed with the RAF rule and fatigue noted. Smooth convergence may

	be assessed with the RAF rule – push-up method. Jump convergence may be assessed by asking the patient to switch fixation between targets held close together within 33 cm and along the anterior–posterior axial plane.
<i>Accommodation</i>	There is a good level of monocular accommodation but when tested binocularly, the accommodative amplitude is reduced (von Noorden <i>et al.</i> 1973).
<i>Binocular function</i>	A reduced positive fusion range is elicited.

Management

Primary convergence insufficiency has been termed functional where conventional orthoptic exercises are usually successful in the treatment of the convergence weakness (Scheiman *et al.* 2011). Correct the refractive error and treat the cause if possible.

Orthoptic treatment: Intensive orthoptic treatment comprising push-up and accommodative targets are the primary treatment for convergence insufficiency (Lavrich 2010). Home-based exercises reduce symptoms while improving near point of convergence and fusional amplitudes (Naqui *et al.* 2003, Serna *et al.* 2011). Recognition of pathological diplopia should be taught if not already appreciated. Binocular convergence is improved with pen convergence exercises. Recognition of physiological diplopia is then taught and the patient should continue to improve binocular convergence using jump pen convergence with appreciation of physiological diplopia. The range of positive fusional convergence is improved with base-out prisms, the synoptophore, dotcards and stereograms with near fixation. Good voluntary convergence is established where possible. Exercises are discontinued for 1 month and if then still asymptomatic with adequate binocular convergence obtained and maintained with ease, the patient is discharged.

Base-in prisms: If unable to start orthoptic exercises due to very reduced convergence, a small prism (approximately half the measurement of the near angle of deviation) may be given to achieve binocularity at a closer distance. Orthoptic exercises may then be commenced and once convergence has begun to improve, the strength of prism may be decreased and eventually removed. Where convergence insufficiency is related to onset of presbyopia, the spectacle lenses may be decentred to offer prismatic effect or a small prism incorporated into the prescription (Lavrich 2010).

Many cases respond to conventional treatment by orthoptic exercises and tend to be primary in nature. Those that do not respond to treatment are often those of secondary nature and may also have poorer levels of convergence and fusional amplitude, or need additional treatment with prisms or surgery (Schwyzer 1978, Wicks 1994, Rowe *et al.* 2001, Stavis *et al.* 2001).

Convergence insufficiency associated with accommodative insufficiency may be treated with plus lenses and base-in prisms.

Medial rectus resection with bifocals may be considered with larger angle deviations (von Noorden 1976, Hermann 1979).

Convergence paralysis

This is a condition in which there is an absence of convergence.

Aetiology

There is usually a history of closed head injury. The condition may be secondary to various organic processes such as encephalitis, diphtheria, multiple sclerosis and occlusive vascular disease involving the rostral midbrain. It may also occur in isolation as a sequence of a viral illness (Kothari *et al.* 2009).

Investigation

<i>Case history</i>	Diplopia is appreciated but often not for distance.
<i>Cover test</i>	Exotropia is noted for near fixation with crossed diplopia.
<i>Accommodation</i>	This is often normal unless the intraocular muscles are involved leading to mydriasis and absent accommodation.
<i>Convergence</i>	There is an absent near point of convergence.
<i>Binocular function</i>	An absent base-out prism fusion range is detected.

Management

The cause of the condition is treated where possible and plus lenses provided for near vision.

Occlusion: This is often the best practical treatment.

Base-in prisms: Diplopia is not appreciated equally at all distances and as a result it is difficult to join with prisms.

Surgery: Unpredictable results are usual. Often, graded surgery is undertaken to account for the greater angle of deviation at near fixation.

Convergence spasm

This is a condition in which there is excessive convergence, most often associated with accommodation spasm, which has been discussed in the previous section for accommodative anomalies.

Specific learning difficulty

Reading difficulties may relate to a number of conditions considered as specific learning difficulties. These include dyslexia and Meares Irlen scotopic sensitivity syndrome.

Those with dyslexia often possess at least average intelligence but have problems processing information and generating output (Handler & Fierson 2011). Handler & Fierson (2011) specifically state that language-based learning disability produces problems with decoding, fluent word recognition, rapid automatic naming and reading comprehension skills. Poor visual coding is reported with dorsal stream deficits (Vidyasagar & Pammer 2010). Abnormalities are also reported in several inter-related brain areas (Brunswick *et al.* 1999).

Meares Irlen syndrome may be diagnosed in individuals sensitive to certain colour wavelengths (Irlen 1994). This sensitivity interferes with visual processing and leads to visual distortion and difficulties with word recognition.

During investigation of patients with specific learning difficulties, the rate of reading test may be undertaken to objectively assess the speed and accuracy of reading. In addition, the developmental eye movement test may be used to assess the horizontal scanning behaviour in a number naming task (Northway 2003). Importantly, any other ocular cause of reading impairment must be excluded by a full orthoptic and ophthalmic assessment. Such causes include decompensating heterophoria or heterotropia, convergence insufficiency, accommodative anomalies, refractive error, poor saccadic tracking and poor binocular vision (Powers *et al.* 2008, Dusek *et al.* 2010, Palomo-Alvarez & Puell 2010). Management should focus on treating any associated ocular cause. Coloured overlays and tinted glasses or contact lenses have been reported to improve rate of reading but with conflicting results as to their efficacy (Evans *et al.* 1994, Speld 1989). Treatment strategies to aid reading may include phonics, reading fluency, vocabulary and comprehension and phonemic awareness (Hatcher *et al.* 2004).

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13 Ptosis and Pupils

Ptosis

The levator muscle is the primary elevator of the upper lid. Its failure to function properly produces a ptosis. Secondary elevators include Müller's muscle and the Frontalis muscle.

Aetiology

Congenital

1. *Isolated simple ptosis*: This may be due to weakness of the levator muscle alone or with ipsilateral superior rectus muscle paresis.
2. *Blepharophimosis*: There are pronounced inverse epicanthic folds, thickening and ptosis of the lids, wide separation of the medial canthi and slit-like palpebral apertures.
3. Congenital III nerve palsy (see Chapter 14).
4. Double elevator palsy (see Chapter 14).
5. Marcus Gunn jaw-winking syndrome.
6. Congenital misdirection syndrome/aberrant regeneration (see Chapter 14).
7. Congenital fibrosis syndrome (see Chapter 15).
8. Periodic ptosis as with cyclic oculomotor palsy (see Chapter 14).
9. Transient neonatal myasthenia gravis (see Chapter 16).
10. Congenital Horner's syndrome.

Acquired

Myogenic (see also Chapter 16)

1. Myasthenia gravis
2. Muscular dystrophy
3. Myotonic dystrophy
4. Chronic progressive external ophthalmoplegia

5. Myositis
6. Senile

Neurogenic (see also Chapter 14)

1. III nerve palsy
2. Aberrant regeneration of III or VII nerve
3. Horner's syndrome
4. Drug induced, for example botulinum toxin

Mechanical (see also Chapter 15)

1. Space-occupying lesions involving the lid
2. Trauma
3. Scar tissue

Differential diagnosis

Ptosis must be differentially diagnosed from pseudoptosis, some causes for which include the following:

1. Ipsilateral hypotropia
2. Ipsilateral enophthalmos
3. Ipsilateral microphthalmos
4. Ipsilateral ptosis bulbi
5. Contralateral proptosis
6. Contralateral lid retraction

Investigation

<i>Case history</i>	Signs of ptosis are often first observed while taking a case history. One should also look for the presence of strabismus, abnormal head posture and pupil abnormalities. The patient is asymptomatic if congenital but with acquired ptosis it may appear worse in the evenings or with fatigue (myasthenia gravis). The patient may appreciate diplopia with associated strabismus and may have blurred vision if mydriasis is present. There may be a history of associated syndromes and systemic disease (Lee <i>et al.</i> 2002), a previous history of facial nerve palsy (Chen <i>et al.</i> 2004) or botulinum toxin injection (Rowe & Noonan 2009).
<i>Refraction</i>	There is an increased frequency of myopia in congenital ptosis (Gusek-Schneider & Martus 2001).
<i>Abnormal head posture</i>	The patient may adopt a chin elevation to compensate for the low lid (McCulloch & Wright 1993).

Visual acuity

Unilateral ptosis causes stimulus deprivation amblyopia depending on the degree of ptosis and age of patient and can occasionally cause anisometropic amblyopia (Anderson & Baumgartner 1980, Dray & Leibovitch 2002, Berry-Brincat & Willshaw 2009). There is a high risk for amblyopia (Oral *et al.* 2010) and the need for head posturing indicates that the visual axis is at least partially obscured (Fiergang *et al.* 1999).

With III nerve palsy, there may be blurred vision due to mydriasis.

Cover test

There is an increased rate of strabismus in eyes with moderate to severe ptosis (Oral *et al.* 2010).

Ocular motility

It is necessary to observe superior rectus function and changes in degree of ptosis in different positions of gaze and with jaw movement. The pupils are examined for signs of miosis or mydriasis. Assessment of lid position and levator function is important and both eyes must always be assessed for comparison. Patients often use frontalis muscle overaction to compensate for ptosis and it is important that this is abolished when assessing the lids. The patient closes the eyes and the examiner presses the brow against the frontal bone to prevent use of the frontalis muscle. The patient then opens the eyes.

The presence or absence, and the height of the skin crease must be noted. The lid crease tends to be higher in cases of congenital ptosis and lower than the other eye in cases of senile ptosis.

When assessing the patient to ensure that the ptosis is truly unilateral and not masked bilateral due to frontalis overaction, frontalis muscle action should be abolished in the apparent ptotic eye and the lid position of the other eye reassessed.

It is important to measure the palpebral aperture of either eye for evaluation of the extent of ptosis. This can be measured with the patient fixing in the primary position and a measurement taken using a rule held against the patient's eyes.

Levator function is assessed by comparing the degree of levator action between upgaze and downgaze. The patient looks down and the upper lid position is measured on a rule held against the face/eye. The patient looks up and the lid position is again measured on the rule (without the rule being moved). The difference is noted and the normal measurement is in the region of 15 mm. A good grade of levator function is recorded for measurements of 12 mm or more, fair grade for measurements of 6–12 mm and poor grade for measurements of less than 5 mm.

The presence of Bell's phenomenon should be assessed and any associated pupil abnormalities or synkinetic movements documented.

<i>Accommodation</i>	Defective accommodation is frequent with III nerve palsy and Horner's syndrome.
<i>Non-orthoptic tests</i>	Ophthalmic, photographs, neurologic, radiologic, muscle biopsy and general medical.

Management

Orthoptic management

Aims of management are to improve visual acuity and eliminate symptoms.

Correct the refractive error and treat amblyopia if present. If unable to assess acuity in young children with obscuration of the visual axis by ptosis, prophylactic part-time occlusion should be considered until acuity can be reliably assessed (Fiergang *et al.* 1999).

- *Observation:* Marcus Gunn syndrome often improves. It is important to monitor the vision of children with unilateral ptosis and monitor the progress of the condition and ptosis for signs of deterioration or recovery.
- Eliminate or join associated diplopia from associated strabismus and/or ocular motility conditions with *prisms or occlusion*.
- Treat the cause of acquired conditions and await any recovery.

Surgical management

Surgery is considered according to the degree of ptosis, the type and cosmesis. Ptosis procedures include brow/frontalis suspension with mersilene or autogenous fascia lata, anterior and posterior levator resection or reinsertion, levator transposition and Fasanella servat with good surgical outcome in most cases (Lee *et al.* 2002).

For a small degree with good levator function, a levator resection or Fasanella servat procedure may be considered. For a moderate degree with good levator function, a levator resection may be considered. For a moderate degree with fair levator function, a large levator resection, tuck or advancement is an option. For a severe degree with poor levator function, a frontalis sling or brow suspension is considered (Yagci & Egrilmez 2003, Morris *et al.* 2008, Chong *et al.* 2010). Use of silicone sling with brow suspension has been reported as effective in allowing adjustment of the sling in cases where the ideal lid position is not achieved after primary surgery (Lamont & Tyers 2010).

Bell's phenomenon and lagophthalmos must be considered when planning surgery as these factors are important protective mechanisms for the surface of the eye (Yoon *et al.* 2008).

Non-surgical management

- Drugs, for example for myasthenia gravis
- Ptosis props (Lane & Collins 1987, Ramasamy *et al.* 2007)
- Magnets (Schreinzer 1980)

Post-operative complications

These may include the following:

- Corneal exposure (Lane & Collin 1987, Lee *et al.* 2002)
- Eye lid too high or too low post-operatively
- Lash eversion and ectropion
- Defective skin crease
- Conjunctival prolapse

Marcus Gunn jaw-winking syndrome

Aetiology

Congenital

The condition is due to abnormal innervation between III and Vc nerves.

Acquired

The acquired form of the condition is rare and often secondary to space-occupying lesions or syphilis where aberrant regeneration has occurred.

Investigation

<i>Refraction</i>	Patients are often anisometropic and astigmatic.
<i>Case history</i>	The condition is usually detected early as it is noticed when the baby is being fed. It is reported as becoming less obvious with time. Familial occurrence is rare although dominant inheritance can occur.
<i>Visual acuity</i>	There is a high incidence of amblyopia.
<i>Cover test</i>	Strabismus is frequently detected and is commonly unilateral. It is commonly an ipsilateral hypo-deviation (Bowyer & Sullivan 2004).
<i>Ocular motility</i>	Unilateral, partial ptosis is present on the affected side, which may be mild to severe (Demirci <i>et al.</i> 2010). The ptosis is variable and may be bilateral. Retraction of the affected upper lid occurs on movement of the lower jaw, particularly when sucking, chewing or speaking. This occurs when the jaw is moved to the opposite side. Retraction and ptosis phases alternate rapidly producing the winking effect. Associated extraocular muscle paresis may occur.
<i>Binocular function</i>	This may be absent or there may be abnormal retinal correspondence depending on the type of manifest deviation. If latent, there is usually good binocular function.

Management

Correct the refractive error if present and treat amblyopia if indicated.

Surgery may be considered for strabismus if a functional result is expected and for cosmetic appearance. Surgery for a significant hypo-deviation should be undertaken before ptosis surgery to determine the effect on lid position (Bowyer & Sullivan 2004). Surgery for ptosis is required if marked. Where possible, ptosis surgery is delayed as much as possible as many cases appear to improve with age. However, if ptosis is marked and covering the pupil, intervention is advisable earlier to prevent amblyopia. Generally, a mild wink and small degree of ptosis responds to unilateral levator surgery whereas for moderate/severe wink and ptosis, bilateral surgery involving levator weakening and brow suspensions should be considered to achieve a symmetrical result (Bowyer & Sullivan 2004, Demirci *et al.* 2010). The most common complications are eyelash ptosis and loss of lid crease (Cates & Tyers 2008).

Lid retraction

Lid retraction may be noted in a number of conditions:

1. Thyroid eye disease.
2. Inferior rectus tethering.
3. Ptosis of the contralateral eye.
4. Midbrain disease.
5. Post-operatively due to overliberal ptosis surgery or surgery involving the superior rectus or inferior rectus.

Management

Botulinum toxin has been used as an option to lower the upper eyelid in cases of lid retraction with good results (Chuenkongkaew 2003, Uddin & Davies 2002). Where a long-term option is required, surgery can include release and excision of tethering and/or scar tissue if relevant, Müller's muscle recession and transection of the upper eyelid (Hintoshchich & Haritoglou 2005).

Pupils

The sphincter pupillae receives parasympathetic nerve supply and constricts the pupil (miosis). This pupillary light reflex is a four-neurone arc. The first-order neurone passes from the retina to the pretectal nucleus. The second neurone passes from the pretectal nucleus to the Edinger-Westphal nucleus and the third neurone passes from there to the ciliary ganglion via the third cranial nerve. The fourth-order neurone passes from the ciliary ganglion to the sphincter pupillae via the short ciliary nerves.

The dilator pupillae receives sympathetic nerve supply and dilates the pupil (mydriasis). This is a three-neurone pathway. The first-order neurone passes from the posterior hypothalamus to the cilio-spinal centre of Budge situated from C8 to T2 in the spinal cord. The second neurone (pre-ganglionic pathway) passes from the spinal cord to the superior cervical ganglion via the vertebral sympathetic chain. The third-order neurone (post-ganglionic pathway) passes from the superior cervical ganglion via the internal carotid artery, through the cavernous sinus to the short posterior ciliary nerves and on to the dilator pupillae.

The near reflex is a triad of miosis, accommodation and convergence. The final pathway of the near reflex is the same as the light reflex. The centre is thought to be in the midbrain in a more ventral position than the light reflex pretectal nucleus.

The accommodative reflex pathway involves neurones passing along the visual pathway to area V1 and then to area 19. Cortical fibres pass via the internal capsule and synapse in the Edinger-Westphal nucleus. Post-synaptic fibres pass along the third cranial nerve to the ciliary ganglion and on to the ciliary body via the long ciliary nerves.

Factors affecting pupil size

These include the following:

- Intensity of retinal illumination
- Near effort reflex
- State of retinal light adaptation
- Supranuclear influences; frontal and occipital cortex
- Reticular formation of brainstem
- Age
- Constant state of movement = hippus

Pupil functions

The functions of the pupils are to:

- regulate amount of light reaching retina;
- reduce chromatic and spherical aberrations produced by peripheral imperfections of cornea and lens;
- increase depth of focus/field.

Classification of pupil anomalies

Unilateral

1. Amaurotic pupil.
2. Anisocoria.

3. *Drugs:* Cycloplegics such as cyclopentolate and atropine will cause mydriasis as may antihistamines and antidepressants. Miosis occurs with drugs such as pilocarpine, phospholine iodide, antipsychotics and narcotics.
4. Holmes–Adie pupil.
5. Horner's syndrome.
6. Internal ophthalmoplegia.
7. Relative afferent pupil defect.
8. *Third nerve palsy:* Mydriasis, if pupil involvement – Hutchinson pupil.
9. Ocular disease such as uveitis, glaucoma, iritis and anterior segment ischaemia results in miosis.
10. Accommodative convergence spasm.
11. Trauma may result in unilateral mydriasis.

Bilateral

1. Accommodative convergence spasm
2. Argyll–Robertson pupil
3. *Drugs:* As above

Investigation

<i>Case history</i>	Pupil abnormalities may be noted while taking a case history. Other signs that may be noted include ptosis, strabismus and heterochromia, gait and balance. History taking may reveal information about management of ocular disease, trauma and issues with general health.
<i>Visual acuity</i>	This may be blurred with mydriasis. An increased depth of focus may be noticed with miosis.
<i>Cover test</i>	Associated strabismus may be seen in cases of pupil involving third nerve palsy. Esotropia is frequently seen with accommodative convergence spasm.
<i>Ocular motility</i>	Extraocular muscle pareses will be noted with associated third nerve palsy. Vertical gaze palsy is commonly noted with dorsal midbrain (Parinaud's) syndrome.
<i>Accommodation</i>	Response to a near target should be noted as normal miosis in response to accommodation and an absent reaction to light stimulus is indicative of light–near dissociation. Accommodation may be defective where mydriasis is noted and an increased depth of focus may be noted with miosis.
<i>Pupil size and reaction</i>	The normal pupils are round and of equal size in either eye. Both react equally to light and accommodation. Pupil size may be irregular due to posterior synechiae from trauma or ocular inflammation. Difference in pupil size (anisocoria) will be noted as either mydriasis of one eye or miosis of the other eye.

The pupillary response to a light stimulus should be noted in either eye and any difference between either eye noted such as an absent or sluggish reaction to light.

When examining the pupils, use a distant fixation to prevent accommodation miosis. General observations should note the speed, quality and symmetry of responses.

Swinging flashlight test

The patient fixates on a distant object that prevents accommodative miosis. A bright light is shone onto the pupil of one eye that should immediately constrict followed by a slow dilation to an intermediate point. This is repeated for the other eye with the light being rapidly transferred from one eye to the other and back several times. A comparison of the pupil reactions will identify the presence of an afferent or efferent pupil defect. The direct reflex occurs in the eye stimulated by the light whereas the consensual (indirect) reflex occurs in the eye unstimulated by the light.

Anisocoria

Anisocoria due to a miosis pupil will show increased anisocoria in dark conditions. Anisocoria due to a dilated pupil shows increased anisocoria in light conditions. Presence of anisocoria generally indicates a defect of the efferent pupillary pathway, which may be physiological and benign or pathological (Wilhelm 2011).

Amaurotic pupil

Defective pupil reflexes are due to damage to the optic nerve and blindness. Both pupils are seen to react with stimulation of the normal eye. No reaction is seen with stimulation of the affected eye. The near-reflex response is normal.

Holmes–Adie pupil

This is a myotonic pupil with absent or diminished tendon reflexes due to a ciliary ganglion lesion (Wilhelm 2008). A dilated pupil is noted with poor direct and consensual reaction to light. There is accommodative paresis and vermiform movements of the iris may be seen on slit-lamp examination. Once the pupil finally constricts, it is slow to dilate in the dark. It occurs mostly in females aged 30–40 years. The diagnosis is confirmed with instillation of either pilocarpine 0.1% or mecholyl 2.5% that will constrict the affected pupil but not the normal pupil. There may be coexistent reduction in knee-jerk reflexes. The pupil defect may be associated with ophthalmoplegic migraine in which resolution of the III nerve palsy occurs but with persistence of the pupil abnormality (Iannetti *et al.* 2009). With treatment, the accommodative response may improve, whereas the light response does not.

Horner's syndrome

There is total or partial interruption of the sympathetic chain to the dilator pupillae with resulting ipsilateral pupil miosis, ptosis, apparent enophthalmos and heterochromia (Wilhelm 2008). Increased amplitude of accommodation may be present. If the lesion is pre-ganglionic, there will also be an associated anhydrosis of the ipsilateral side of the face. Central causes of the syndrome include hypothalamic or brainstem lesions (vascular, tumour, multiple sclerosis). Pre-ganglionic causes include tumours in the chest cavity, neck lesions (trauma, abscess, thyroid gland tumour) and neck or thyroid gland surgery. Post-ganglionic causes include superior cervical ganglion lesions (trauma or surgery), internal carotid artery lesion (dissection, trauma, arteritis, tumour compression or migraine) and cavernous sinus lesion (tumour, inflammation, fistula or thrombosis). Paediatric Horner's syndrome is due to birth, surgical or other trauma in 65% of cases (Smith *et al.* 2010) but rarely with an underlying mass lesion. The diagnosis is confirmed with instillation of cocaine 4% in either eye that will dilate the normal eye but not the affected pupil. For differential diagnosis of pre- and post-ganglionic lesions, hydroxyamphetamine 1% (paredrine) is instilled that dilates the affected pupil if there is a pre-ganglionic lesion but not if post-ganglionic.

Ross syndrome

This syndrome consists of a tonic pupil, hypo reflexes, segmental anhidrosis and heat intolerance. It is due to post-ganglionic autonomic abnormalities in the central nervous system (Hedges & Gerner 1975).

Hutchinson pupil

This is a dilated pupil in the presence of a third nerve palsy. It is reported in 38% of third nerve palsies due to ischaemic lesions and 80% of third nerve palsies due to aneurysm (Kupersmith *et al.* 2006). Thus, the presence of a dilated pupil along with third nerve palsy requires detailed angiography imaging.

Internal ophthalmoplegia

This is due to a lesion in the efferent parasympathetic pupillary pathway that causes loss of direct and consensual light reflexes and loss of miosis to a near stimulus on the affected side.

Relative afferent pupillary defect

This is caused by an optic nerve lesion that is not severe enough to cause an absence of light perception. The affected pupil does not constrict to direct light stimulation but does react on consensual testing. The pupil of the affected eye will be seen to dilate as the light is transferred to it on the swinging flashlight test.

Argyll–Robertson pupil

A lesion in the rostral midbrain affects the light reflex fibres to the Edinger–Westphal nucleus (Thompson & Kardon 2006). This results in miotic irregular pupils and light–near dissociation. Both pupils are involved but often to an asymmetrical degree. Causes of the condition include neurosyphilis, diabetes and multiple sclerosis.

Springing pupil

This clinical sign involves pupil dilation associated with migraine (Hallett & Cogan 1970) and can be linked with a Holmes–Adie pupil (Miller *et al.* 2010). Subtle sympathetic and parasympathetic pupil abnormalities are reported to persist in migraine (Harle *et al.* 2005).

Midbrain correctopia

In this pupil condition, an eccentric or oval pupil is seen due to rostral midbrain disease (Selhorst *et al.* 1976, Shams *et al.* 2010, Wilhelm 2011).

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14 Neurogenic Disorders

This chapter discusses the neurogenic disorders including cranial nerve palsies, isolated extraocular muscle pareses and the ophthalmoplegic syndromes. A palsy indicates loss of nerve function. This may be complete (no function) or partial (some residual function). The term paresis also applies to a partial palsy.

Cranial nerve palsies may be caused by a variety of aetiologies including vascular, trauma, neoplasm, space-occupying lesion, aneurysm, inflammation and infection, iatrogenic, systemic disease or unknown cause (Rucker 1958, 1966, Rush & Younge 1981, Richards *et al.* 1992, Tiffn *et al.* 1996, Park *et al.* 2008). Sixth nerve palsy is more common than fourth and third nerve palsies. Approximately 50% show spontaneous recovery, with the best recovery often reported for vascular aetiologies.

III (third) cranial nerve

The III nerve divides into two branches. The superior branch supplies the levator palpebrae superioris muscle and the superior rectus. The inferior branch supplies the medial rectus, inferior rectus and inferior oblique muscles. A complete lesion of the III nerve involving both branches will result in a deficit of elevation, adduction and depression in abduction. There will be an accompanying ptosis and pupil dilatation.

Alternatively, there may be a lesion affecting only one or other of the branches and these lesions may be partial or complete.

Aetiology

Localisation of lesion

Nuclear

Nuclear III often produces bilateral defects of ocular motility and lid function. The levator palpebral superioris muscles share a common central nucleus that produces

bilateral ptosis if affected (Pratt *et al.* 1995). The superior rectus muscle has contralateral innervations. Rostral nuclear lesions may result in pupil involvement with spared lid function whereas caudal nuclear lesions may result in bilateral ptosis (Bioussé & Newman 2000, Saeki *et al.* 2000). A nuclear lesion may also result in the following:

- Unilateral III with bilateral ptosis
- Unilateral III with contralateral superior rectus underaction
- Isolated extraocular muscle palsy of inferior rectus, inferior oblique or medial rectus muscles (Brown 1957)
- Bilateral III with spared levator function

Internuclear

- Internuclear ophthalmoplegia
- *Weber's syndrome*: III and contralateral hemiplegia due to lesion of corticospinal tract
- *Benedikt's syndrome*: III and contralateral ataxia and intension tremor
- *Claude's syndrome*: Lesion of the red nucleus and III nucleus producing an ipsilateral III nerve palsy and contralateral ataxia (Broadley *et al.* 2001)

Infranuclear

This is due to a lesion along the nerve pathway. Third nerve palsy may be central, sparing the pupil or peripheral with pupil involvement. If the pupil is spared, the cause is most likely vascular. When the pupil is involved, the cause is likely to be an aneurysm (Goldstein & Cogan 1960). However, pupil sparing in children (unlike in adults) may not be helpful in differentiating the causes of the palsy and third nerve palsy in children warrants full neurological investigation (Ng & Lyons 2005). The two most common causes are the following:

- *Aneurysms in the Circle of Willis*: Those involving the posterior communicating artery are particularly associated. There is often pupil involvement resulting in dilatation.
- *Diabetes*: Third nerve lesions associated with underlying vascular disorders are often pupil sparing.

Trauma, cavernous sinus lesions and space-occupying solid lesions are other major factors that may result in III nerve palsies (Rush & Younge 1981, Tiffin *et al.* 1996, Rowe & Eariss 1999). Trauma accounts for a small percentage of acquired III nerve palsy (Richards *et al.* 1992). Space-occupying lesions involving the uncal portion of the temporal lobe can cause uncal herniation with compression of the III nerve. Cavernous sinus lesions frequently involve other cranial nerves in addition to the III and lesions may include bacterial and fungal inflammation, sarcoid, Wegener's granulomatosis, Tolosa-Hunt Syndrome, internal carotid artery aneurysm or fistula.

Typical III nerve palsy

Investigation

Visual acuity It is necessary to lift the ptotic lid to evaluate visual acuity. This may be reduced due to mydriasis, particularly for near visual acuity.

Cover test An exo- and hypo-deviation is present.

Ocular motility The superior branch supplies the levator muscle and superior rectus muscle. The inferior branch supplies the medial rectus, inferior rectus and inferior oblique muscles and carries the pupil reflex fibres. There will be limited elevation, depression and adduction, which may be complete or partial limitations, depending on the extent of paresis/palsy. The examiner should always check for the presence of IV nerve function by asking the patient to attempt to look down and outwards and observe for the presence of incyclo-torsion during this movement.

There is likely to be a ptosis present in which case the upper lid will have to be raised by the examiner (or an assistant) in order to perform the ocular motility assessment. The pupil may be dilated, depending on the underlying pathology (Fig. 14.1).

It is important to check for unilateral versus bilateral signs and extent of limitations (ductions) versus version movements.

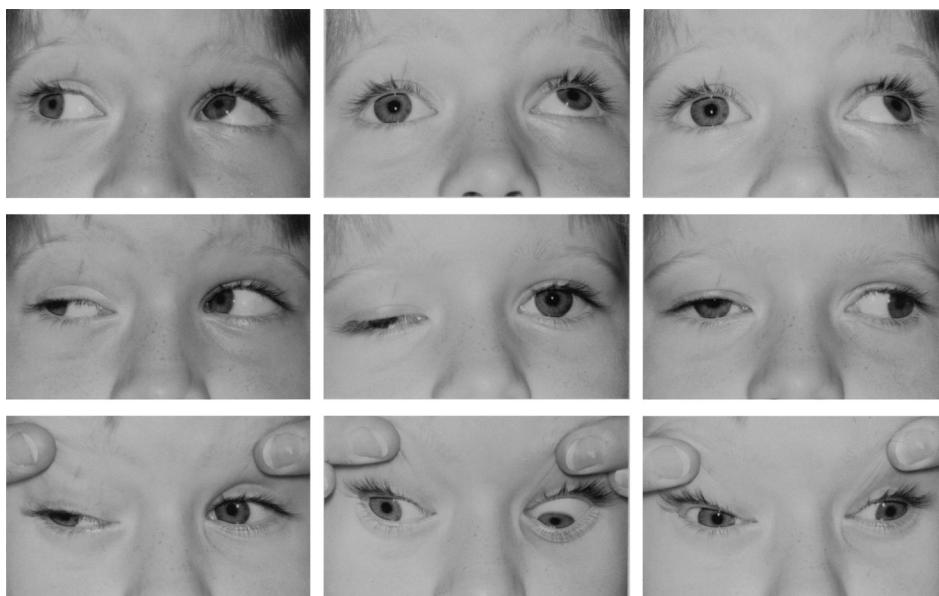


Figure 14.1 Right III nerve palsy.

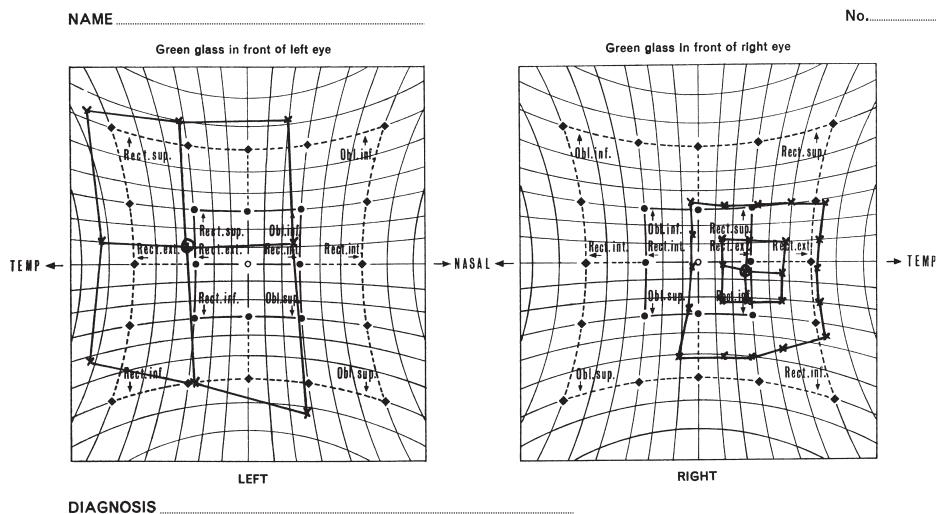


Figure 14.2 Hess chart of right III nerve palsy.

Convergence

This will be absent if the medial rectus muscle is paralysed. This is usually absent unless the III nerve paresis is mild and partial.

Accommodation

If the underlying cause of the lesion has resulted in pupillary dilatation, then fibres to the ciliary body are also likely to be involved so that accommodation will be defective.

Hess chart

The affected eye will show a markedly constricted field whereas the other eye demonstrates overaction of its muscles (Fig. 14.2).

Diplopia

There will be constant diplopia unless complete ptosis is present and blocks the vision of the affected eye.

Aberrant regeneration

This is a change in the actions of muscles supplied by the third nerve due to regrowth of damaged nerve fibres following complete or severe third nerve palsy (Shuttleworth *et al.* 1998). It is liable to occur when either trauma or an aneurysm has caused the lesion (Cox *et al.* 1979, Rossillion *et al.* 2001). Aberrant regeneration may occur from weeks to months after the onset of the III nerve paresis. The factors underlying aberrant regeneration are unclear but may involve either a central or peripheral response. In the former, there is a mass response of the damaged nerve resulting in growth to the wrong muscle. In the latter, abnormal growth of damaged axons may occur locally. Aberrant regeneration may also occur between the III and V nerves with abnormalities of intraorbital structures and central nervous system pathways following trauma (Buckley *et al.* 2005).

Investigation

<i>Cover test</i>	Combined horizontal and vertical deviations are common.
<i>Ocular motility</i>	Abnormal movements often involve the eyelids and pupils. There may be horizontal gaze and lid synkinesis (Chua <i>et al.</i> 2000) and limited elevation and depression with globe retraction on attempted vertical movements or ipsilateral adduction on attempted elevation or depression.
<i>Ptosis</i>	The lid may rise on attempted depression, adduction and occasionally abduction (pseudo-Graefe phenomenon).
<i>Pupil</i>	This may constrict on attempted adduction, elevation and depression (pseudo-Argyll Robertson pupil).
<i>Convergence</i>	This may occur on attempted elevation.

Cyclic oculomotor palsy

This rare condition is usually congenital and unilateral in origin (Burian & Van Allen 1963). It is often associated with a partial III nerve palsy with some degree of ptosis. Acquired cyclic ocular motor palsy may occur following irradiation of the skull base and is similar to ocular neuromyotonia (Miller & Lee 2004). It may also be due to a compressive lesion (Bateman & Saunders 1983). The condition is described as having cyclical fluctuation in two phases:

1. *Paralytic phase:* There is a partial III nerve palsy.
2. *Miotic phase:* There is convergence, lid retraction, accommodation and pupil constriction.

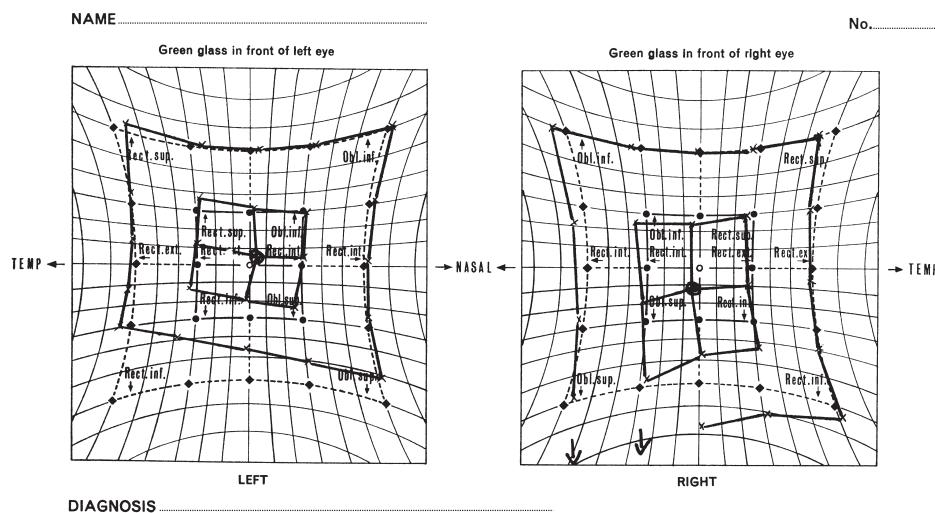


Figure 14.3 Hess chart of left inferior rectus palsy.

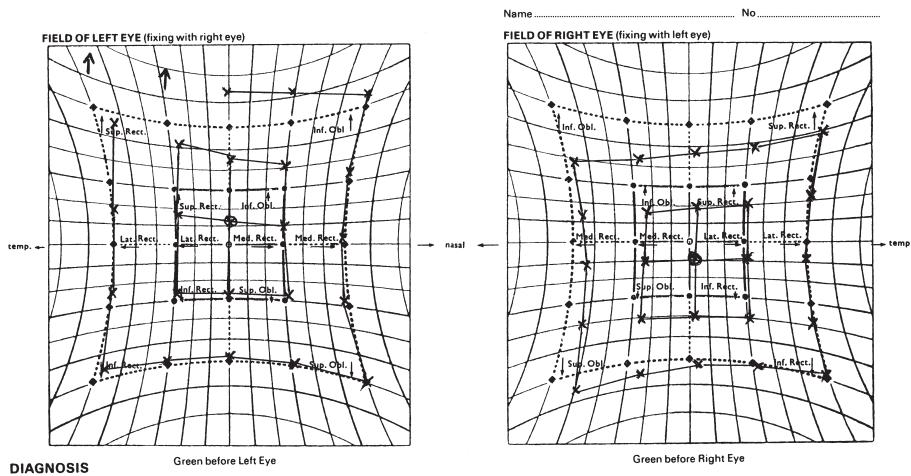


Figure 14.4 Hess chart of right inferior oblique palsy.

Single muscle palsies

The following isolated muscle palsies are all recognised.

1. *Medial rectus*: This produces an exo-deviation, which is greater for near fixation.
2. *Inferior rectus*: This produces hyper- and exo-deviation (Fig. 14.3).
3. *Superior rectus*: This is often bilateral and may present with a V exo pattern.
4. *Inferior oblique*: This is a feature of an A eso pattern (Fig. 14.4).

Differential diagnosis of single muscle palsies

The following conditions should be considered:

Medial rectus palsy	Atypical Duane's retraction syndrome Unilateral/bilateral internuclear ophthalmoplegia Myogenic (myasthenia gravis) Mechanical limitation (thyroid eye disease) Trauma (blowout fracture)
Inferior rectus palsy	Brown's syndrome Trauma (blowout fracture)
Inferior oblique palsy	Mechanical limitation (thyroid eye disease)
Superior rectus palsy	

Double elevator palsy

This often has a congenital origin and is presumed to be caused by a supranuclear defect. The superior rectus and inferior oblique muscles of the same eye are affected (Jampel & Fells 1968, Strachan & Innes 1987).

Investigation

<i>Cover test</i>	There is a hypo-deviation in the primary position that may be manifest or latent.
<i>Ocular motility</i>	There is limited elevation of one eye in both adduction and abduction. Ptosis and/or pseudoptosis may be present. Bell's phenomenon is usually present.
<i>Abnormal head posture</i>	The chin is elevated to compensate for the palsy.
<i>Binocular function</i>	If there is only a small angle of deviation in the primary position, this may be controlled with or without an abnormal head posture, resulting in a latent hypo-deviation. In these circumstances, binocular single vision is present.
<i>Forced duction test</i>	There may be full passive movement (negative result) or it may be positive, depending on cause that is important for surgical planning.

Differential diagnosis double elevator palsy

The following conditions should be differentiated from double elevator palsy as they will have a positive forced duction test:

- Blowout fracture
- Thyroid eye disease
- Brown's syndrome
- Congenital fibrosis of the inferior rectus muscle
- General fibrosis syndrome

Conditions with a negative forced duction test, which should be included in the differential diagnosis, are anomalous superior or inferior rectus muscle insertions.

Management of third nerve palsy

In children, any associated amblyopia should be treated. Any underlying cause for the lesion should be investigated and treated. As with most conditions affecting the extraocular muscles, it is important to allow a period for recovery of any muscle function and then wait until stability of this has been demonstrated. Recovery is more common in vascular aetiologies (Rowe & Eariss 1999).

Prisms may be helpful with partial III nerve palsy where there is a small deviation in primary position that can be joined with prisms. Long-term prisms may be incorporated into a spectacle correction.

Occlusion is indicated for diplopia, which cannot be joined by prisms because of a complete palsy and incomitant ocular motility and where surgery is not (yet) an option. Forms of occlusion include a total occlusive patch, frosted lenses, Bangerter filters or an occlusive contact lens.

Surgery is indicated to correct the strabismus and any associated ptosis. Squints are treated initially as the ptosis acts as a mechanical barrier to diplopia. However,

if the action of raising the lid has an effect on the strabismus, this must be accounted for in the surgery undertaken.

In the case of a complete palsy, there is a poor prognosis for outcome and the aim of surgery is to improve cosmesis plus, if possible, to achieve some field of binocular single vision. Any ptosis is subsequently corrected. It should be noted that an adequate Bell's phenomenon is needed before ptosis can be repaired.

Strabismus surgery for complete palsies is usually performed in stages. The level of function of each involved muscle must be carefully evaluated and the presence of secondary ipsilateral lateral rectus contracture identified. All patients require evaluation for possible disruption of central fusion. If serious neurological disease is found, treatment may need to be modified in light of the patient's general condition.

Total third nerve palsy

A very large maximal lateral rectus recession (on hang back sutures) to totally abolish abduction is undertaken. Traction sutures may be used to anchor the eye in its new position of gaze to further enhance the result whereby non-dissolvable sutures are passed through the upper lid and anchored on bolsters. These are left in place for a minimum 6-week period with improvements of approximately two-third of the pre-operative angle (Khaier *et al.* 2008). Superior oblique tendon resection and transposition to the area between the medial rectus and superior rectus insertions may create a chronic adducting force that will ensure long-term stability (Peter 1934, Metz & Yee 1973, Saunders & Rogers 1982, Maruo *et al.* 1996, Young *et al.* 2000, Villasenor *et al.* 2000).

Where aberrant regeneration has occurred, surgery may be aimed at placing the eyes in a position that takes into account any improved lid function, thus stimulating the levator muscle and improving ptosis (Parulekar & Elston 2003).

Partial third nerve paresis

Where there is isolated muscle involvement, the paretic muscle with residual function is resected and the antagonist is recessed, preferably with adjustable sutures. Where there is multiple muscle involvement, resection of the paretic muscles with recession of the lateral rectus is advised. The insertions may be transposed down with absent inferior rectus function to provide a downward force (Metz 1988, Metz 1993). Recession or Faden to the contralateral inferior rectus to match a deficient field can be considered. A Knapp's procedure for inferior division third nerve palsy is a useful option.

Botulinum toxin to the lateral rectus may aid horizontal alignment (Marsh 2008). The patient may then adopt a vertical abnormal head posture to attain fusion (Metz 1988).

In cases of double elevator palsy, a Knapp's procedure may be indicated (Knapp 1969, Burke *et al.* 1992). In this operation, the lateral and medial recti are transposed to the superior rectus insertion. An augmented Knapp's procedure with posterior fixation sutures has been shown to be effective (Snir *et al.* 2005). If the inferior rectus muscle in the affected eye is limited with a positive forced duction

test, then this muscle may be recessed. Transposition surgery alone with negative forced duction test or combined with inferior rectus recession with a positive forced duction test is reported as effective and can be combined with horizontal muscle surgery where there is coexistent horizontal deviation (Yurdakul *et al.* 2009). Ptosis surgery may or may not be required depending on the lid position once ocular alignment has been achieved.

IV (fourth) cranial nerve

The IV cranial nerve supplies the superior oblique muscle only. Any lesion affecting the nerve may result in difficulties of depression, incyclorotation and abduction of the eye.

Aetiology

Location of lesion

Fourth nerve palsy may be due to lesions in the nucleus or fascicular lesions of the midbrain. It can be difficult to differentially diagnose nuclear and fascicular lesions as the IV nerves decussate immediately after exiting the nuclei and exit the dorsal midbrain after a very short intra-midbrain course. The IV nerve is not only susceptible to damage as it exits the midbrain but also is vulnerable in the cavernous sinus and orbital apex.

The condition may be congenital or acquired. The latter group may be caused by the following:

- **Trauma:** Particularly bilateral cases, as the IV cranial nerve is the only nerve to arise from the dorsal surface of the midbrain, and therefore follows a long, winding route that renders it susceptible to injury.
- **Vascular,** for example hypertension.
- **Diabetes.**
- **Space-occupying lesions.**

In congenital superior oblique palsy, the tendon is usually lax and abnormally long (Plager 1990, Plager 1992).

Investigation

Case history In congenital cases, the patients are often affected bilaterally. They may present with any of the following:

- Manifest strabismus without binocular function
- Binocular function, but there may be an abnormal head posture, depending on the extent of paresis

- Symptoms of decompensation that occur when the patient has been binocular, possibly with an abnormal head posture, but it has become too difficult to control the deviation. The patient may suffer from symptoms of diplopia, headaches and asthenopia.

These effects are experienced particularly by members of the older age group and therefore it is necessary to establish the underlying congenital nature of the disorder from an acquired cause.

There is often facial asymmetry in congenital superior oblique palsy consisting of shallowing of the midfacial region between the lateral canthus and the edge of the mouth (Parks 1958). This is found in three-fourth of patients with congenital palsy (Wilson & Hoxie 1993, Paysee *et al.* 1995).

In acquired cases, the paresis may be unilateral or bilateral. The latter is often associated with major closed head trauma. Symptoms of cyclovertical diplopia are experienced, which worsen on downgaze. The presence of an abnormal head posture tends to indicate more long-standing deviations.

Abnormal head posture

In unilateral IV nerve paresis, the patient may present with chin depression with face turn and head tilt away from the affected side. Bilateral cases demonstrate chin depression but there may well be no face turn or head tilt unless one side is affected more than the other.

Persistent abnormal head posture following surgery for IV nerve palsy should be investigated to exclude sternocleidomastoid muscle tightness that may have developed in congenital cases (Lau *et al.* 2009).

Cover test

The test is performed with and without the abnormal head posture for comparison. A latent deviation exists if a compensatory abnormal head posture is adopted. When the test is repeated with the head straight, the deviation will increase and may become manifest.

The affected eye shows a hyper-deviation and an associated eso-deviation. (However, an exo-deviation may be seen if this was present before the onset of the palsy.) It is useful to note that the vertical deviation is commonly greater on near testing than for distance. In bilateral cases, the main deviation usually tends to be a small-angle horizontal deviation.

Ocular motility

The primary underaction of the affected superior oblique muscle results in a number of sequelae. There is overaction of the contralateral inferior rectus and of the ipsilateral inferior oblique muscles. The contralateral superior rectus shows secondary underaction. The combination of these muscle effects is variable and often depends on whether or not the deviation is long standing (Fig. 14.5).

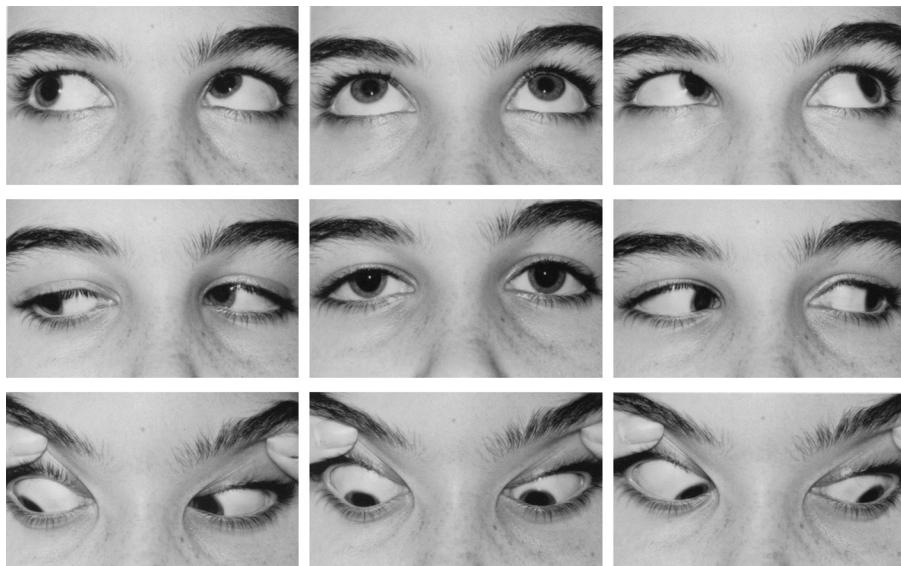


Figure 14.5 Left IV nerve palsy.

In bilateral cases, a V pattern is often present. In the presence of this, one should always suspect a bilateral case, especially where there is an excyclotorsion of greater than 7° and a tendency towards reversal of any hyper-deviation or diplopic images on lateral gaze (Hermann 1981).

Knapp's classification (1974) takes into account the position of maximum deviation in addition to the deviation in primary position:

- I: Greatest deviation is with ipsilateral eye on elevation in adduction due to inferior oblique overaction.
- II: Greatest deviation is with ipsilateral eye on depression in adduction due to superior oblique underaction.
- III: Greatest deviation in all contralateral positions of gaze.
- IV: Greatest deviation in all contralateral positions of gaze and in all downward gaze positions.
- V: Greatest deviation in all downward gaze positions.
- VI: V esotropia, cyclotropia and bilateral superior oblique palsies.
- VII: Poor elevation and depression in adduction of ipsilateral eye due to direct injury to the superior oblique muscle resulting in restriction and paresis.

Convergence

This may be reduced, either due to convergence insufficiency or the vertical deviation. Differentiation is achieved by correcting the deviation and seeing if convergence improves.

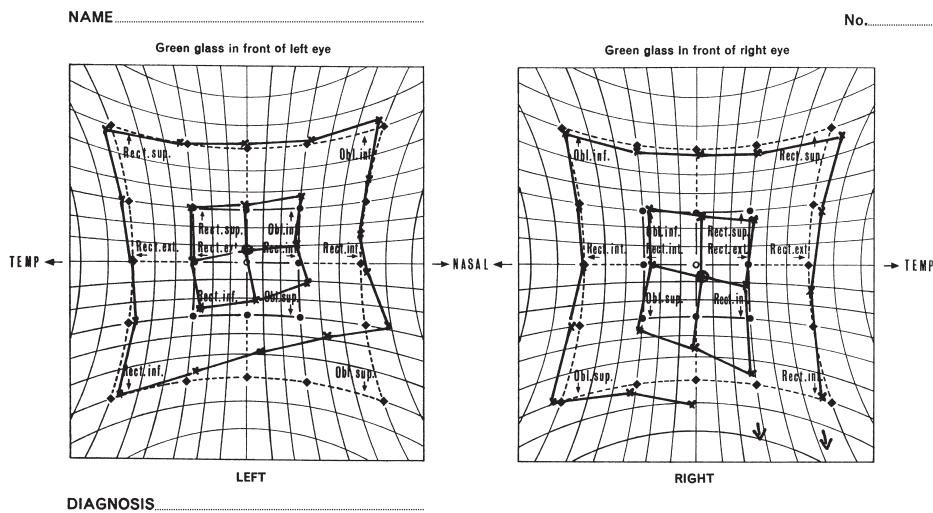


Figure 14.6 Hess chart of left IV nerve palsy.

Binocular function

Assessment of binocular function is carried out with and without the abnormal head posture, if one is present. Binocular single vision cannot be maintained if there is a significant degree of torsion and therefore is often not present in acquired bilateral palsies.

Patients who maintain binocularity tend to have a small angle of deviation associated with a congenital palsy. Some patients with long-standing acquired palsies can maintain binocularity if there is a good fusional control with an extended vertical fusion range. In addition, there may be an extended cyclorotational fusion range. There is often a discrepancy between the subjective report of torsion and the objective measurement of head tilt, cyclo-deviation and retinal rotation of the macula (Ruttum & von Noorden 1983, Jampel & Shi 2002). Cyclofusional reserves can allow for 8° of sensory cyclofusion and 7° of motor cyclofusion.

Hess chart

The Hess chart provides a useful record for the notes and demonstrates the effects of the paresis on the other extraocular muscles (Fig. 14.6).

Field of binocular single vision

The area in which binocular single vision is retained is displaced upwards to the affected side (Fig. 14.7).

Diplopia

The patient experiences a greater degree of diplopia on near testing when looking down. The diplopia is vertical and usually uncrossed. However, if an exo-deviation is present, the horizontal element of diplopia will be crossed.

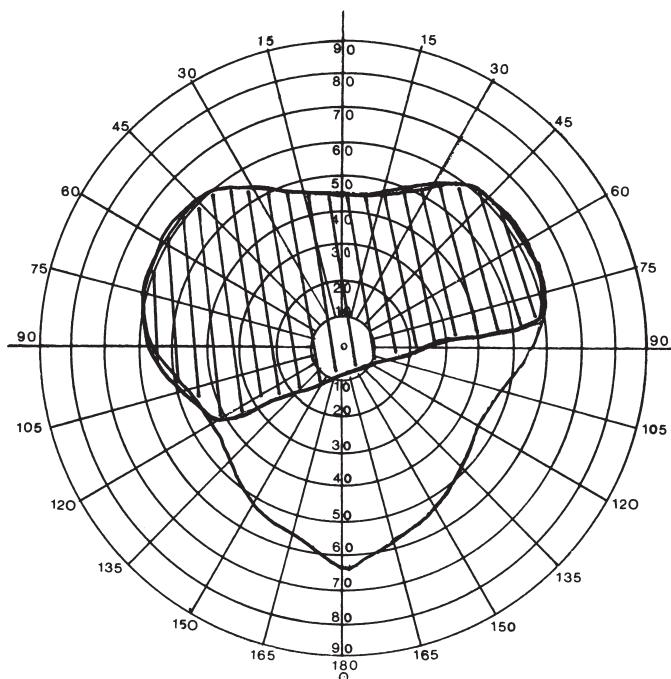


Figure 14.7 Field of binocular single vision of left IV nerve palsy.

Bielchowsky head tilt test

A cover test in the primary position with the head straight will show a hyper-deviation in the affected eye. If there is a positive Bielschowsky head tilt test, on tilting the head towards the affected side, this vertical deviation increases. Tilting towards the unaffected side will show a decrease, which is indicative of a IV nerve palsy. The classic head tilt test is not specific for determining true versus masquerading IV nerve palsy (Demer *et al.* 2011).

Torsion

Diagnostic prisms

Excyclotorsion is frequently present in acquired bilateral cases. Fresnel prisms may be used temporarily to correct the angle of deviation. If the prism alleviates the need for an abnormal head posture, then the indication is that the IV nerve was responsible for the abnormal head posture rather than a non-ocular cause.

Differential diagnosis

Differential diagnosis should be made for the following conditions: thyroid eye disease, ocular surgery, orbital fracture, neurosurgery, childhood strabismus, skew deviation, third nerve palsy, myasthenia gravis and decompensated hyperphoria (Tamhankar *et al.* 2011). Differential diagnosis with skew deviation is by the presence of torsion. Typically, excyclotorsion is seen in IV nerve palsy and incyclotorsion is seen in skew deviation. Furthermore, the deviation in IV nerve palsy remains when the patient is lying down whereas skew deviation resolves on lying down (Wong 2010).

Table 14.1 Differences of superior oblique and superior rectus palsy.

Investigation	Superior oblique	Superior rectus
<i>Cover test</i>	Hyper-deviation if fixing with unaffected eye Deviation greater for near	Hypo-deviation if fixing with unaffected eye Deviation greater for distance
<i>Abnormal head posture</i>	Chin depression	Chin elevation
<i>Ocular motility</i>	Increase in angle on depression	Increase in angle on elevation
<i>Hess chart</i>	Greatest negative displacement on depression	Greatest negative on elevation
<i>Extorsion</i>	Very common	Rare
<i>Bielschowsky head tilt test</i>	Often positive	Usually negative

Table 14.2 Differences of unilateral and bilateral superior oblique palsy.

Investigation	Unilateral	Bilateral
<i>Cover test</i>	Hyper-deviation in primary position reflects extent of palsy	Often only slight hyper-deviation in primary position
<i>Ocular motility</i>	No reversal of hypertropia or and diplopia on lateral versions Slight V pattern may be noted	Reversal of hyper-deviation diplopia on lateral versions Large V pattern
<i>Abnormal head posture</i>	Chin depression, head tilt and head turn	Chin depression
<i>Torsion</i>	Slight extorsion	Extorsion $> 10^\circ$
<i>Bielschowsky head tilt test</i>	Positive with head tilt to affected side	Positive with head tilt to either shoulder

Table 14.1 indicates the differences to be found on examination between a superior oblique palsy and a contralateral superior rectus palsy in long-standing cases.

Unilateral and bilateral cases of superior oblique palsy show a number of different examination results, which are outlined in Table 14.2.

Management

Congenital

Correct any refractive error if present and treat amblyopia if indicated. Surgery may be required for any of the following:

- Strabismus
- Marked abnormal head posture
- Decompensating cases that show moderate/large-angle deviations

Abnormal superior oblique tendons are found in many cases of congenital IV nerve palsy and these cases may respond to a superior oblique tuck procedure (Helveston *et al.* 1981, 1992). Further procedures include weakening of overacting muscles including the inferior oblique and inferior rectus.

Acquired

Treatment of the underlying cause of the IV nerve palsy, for example hypertension. Many cases recover within 3–6 months of onset.

Prisms: These can be used for small angles of deviation. Prisms of up to 10 prism dioptre strength can be incorporated in each lens, depending on the patient's spectacle prescription. Prism incorporation occurs at the stage where the condition is considered to be stable usually with static reports for a minimum of 6 months.

Occlusion: This eliminates diplopia whilst waiting for static reports prior to further treatment.

Botulinum toxin: This injection of the inferior oblique muscle has been utilised to reduce the hypertropic deviation to alleviate symptoms during the recovery period (Bagheri & Eshaghi 2006).

Surgery: Once the degree of recovery is considered to be stable and static (usually a period of 6 months minimum is required for this), surgery may be undertaken. Knapp (1974, 1976) and Scott & Kraft (1986a, 1986b) emphasised the importance of the relative magnitude of hyper-deviation in the various fields of gaze. The fields of greatest deviation should be matched to the muscles that exert their strongest action in those fields. For example, if the inferior oblique muscles overact, this should be recessed. If the inferior oblique does not overact and the ipsilateral superior rectus is restricted, the superior rectus should be recessed. If the superior oblique is lax, it should be tucked.

Where the deviation measures more than 15 prism dioptres, surgery on two or more muscles is required: ipsilateral inferior oblique recession plus ipsilateral superior rectus recession if restricted or ipsilateral superior oblique tuck if lax or contralateral inferior rectus recession or combination as necessary.

In cases with combined horizontal and vertical angles, it is recommended that combined horizontal and vertical muscle surgery be undertaken where the horizontal angle exceeds 8 prism dioptres (Telander *et al.* 2011).

Generally, for large-angle deviations with/without torsion, surgical procedures include the following:

- Weaken the overacting muscles:
 - ipsilateral inferior oblique recession
 - contralateral inferior rectus recession

Adjustable sutures may be used.

- For torsion, a Harada–Ito (Fell's modification) can be performed (Harada & Ito 1964). In this procedure, the anterior portion of the affected superior oblique tendon is placed further anteriorly and laterally. This increases the sagittal axis

on the globe and leads to a greater amount of incyclotorsion compared to depression as the muscle functions. A further modification of the Harada–Ito procedure involves separation of the anterior half of the superior oblique tendon without detachment of the insertion and pulling this half forward and laterally to be sutured to the sclera temporal to the superior rectus insertion (Ehrt & Boergen 2004).

- In bilateral cases, Harada–Ito procedures may be carried out on both sides where there is torsion. If there is no torsion, bilateral inferior oblique recessions may suffice.

Sometimes, a combination of these two procedures is required.

Superior oblique myokymia

This very rare benign, self-limiting disorder is found unilaterally and may follow an acquired superior oblique palsy.

The condition is characterised by the following features:

- The patient suffers recurrent episodes of involuntary rapid eye movements, which may be vertical or torsional. These movements are clearly visible when observed through a slit lamp or direct ophthalmoscope.
- These movements are experienced by the patient with the production of intermittent oscillopsia.
- The patient may also experience torsional monocular diplopia and oscillopsia.

The cause is usually unknown but it has been suggested that it occurs following an alteration in membrane threshold of the IV nerve nucleus resulting in uncontrolled contractions of the superior oblique muscle. These may be due to defective supranuclear input with regeneration of motor neurones in the IV nucleus (Yousry *et al.* 2002, Hashimoto *et al.* 2004) or due to nerve compression by blood vessels in the posterior fossa (Bringewald 1983, Scharwey *et al.* 2000). It is generally a benign condition with a natural history of remitting and relapsing episodes varying up to 28 years (Palmer & Shults 1984, Brazis *et al.* 1994, Kattah & Fitzgibbon 2003). However, occasional associations have been made with the presence of multiple sclerosis, space-occupying lesion and dural arterio-venous fistula (Morrow *et al.* 1990, Geis *et al.* 1996) and it is thus important to undertake neuroimaging to exclude neurological and vascular compression (Webster & Leslie 2004). The patient's symptoms sometimes reduce with time and episodes are often brief, occurring at irregular intervals. There may be long periods of remission, which may last for several years (Kommerell & Schaubele 1980).

Management

Carbamazepine (Tegretol) has been used with some success (Brazis *et al.* 1994). The therapeutic effect is probably related to an elevation of the neural threshold of

the IV nerve to repeated stimulation. The patients need to be followed-up at regular intervals with full blood counts and liver function tests due to possible unwanted effects of bone marrow suppression, aplastic anaemia and hepatic toxicity. Other medical drugs have been used with varying success including propranolol and gabapentin (Leigh & Tomsak 2003). Surgery may be considered in severe cases but is rarely associated with a complete cure. The procedure involves a superior oblique intrasheath tenotomy or nasal tenectomy combined with an ipsilateral inferior oblique recession or myectomy (Staudenmaier 1986, Brazis *et al.* 1994, Agarwal & Kushner 2009) or modified Harada–Ito procedures (Kosmorsky *et al.* 1995). Further surgery after failed primary treatment may include superior oblique myectomy combined with resection of the trochlea (Ruttum & Harris 2009).

VI (sixth) cranial nerve

The VI cranial nerve supplies the lateral rectus muscle only. A lesion affecting the nerve will result in defective abduction of the eye.

Aetiology

The commonly recognised causes of sixth nerve palsy include space-occupying lesions, trauma, vascular insults and inflammation. Sixth nerve palsy secondary to raised intracranial pressure is also commonly regarded as a typical false localising sign. Other causes have included pseudotumor cerebri (idiopathic intracranial hypertension), post-operative complications, viral infection, multiple sclerosis and otitis.

Congenital

- Following birth trauma
- Hereditary
- Infection (maternal)
- Failure of lateral rectus development

Acquired

This differs according to age:

- *Children* (Afifi *et al.* 1992):
 - Space-occupying lesions
 - Infections, bacterial or viral^{a,b}
 - Trauma

^a Gradinego's syndrome is the association of a VI nerve palsy with middle ear infection that has spread to the VI nerve at the petrous-temporal bone.

^b A diagnosis of benign sixth nerve palsy often follows infection, viral illness, immunisation or other such causes in children with good recovery but occasional recurrence (Sturm & Schoffler 2010, Werner *et al.* 1983).

- Raised intracranial pressure
- Decompensated esophoria
- Infantile esotropia with cross-fixation
- Möbius syndrome
- Duane's retraction syndrome
- *Young adults:*
 - Trauma
 - Space-occupying lesions
 - Post-viral inflammation
 - Multiple sclerosis
 - Diabetes
 - High myopia
 - Ophthalmoplegic migraine
- *Older adults:*
 - Vascular
 - Diabetes
 - Space-occupying lesions
 - Senile lateral rectus weakness

Investigation

<i>Case history</i>	There may be a significant medical history as indicated above. Birth history may also be significant. The patient may complain of horizontal diplopia, which is worse in the distance and/or on lateral gaze and relatives or friends may notice the presence of squint (esotropia) or an abnormal head posture.
<i>Visual acuity</i>	This may be reduced if the affected eye fails to fixate properly due to the presence of a marked deviation. Amblyopia will develop in children with unilateral strabismus.
<i>Abnormal head posture</i>	The face is turned towards the affected side.
<i>Cover test</i>	An eso-deviation is present that is often greater on distance testing. The test should be carried out with and without an abnormal head posture, if one is present. The deviation increases without the head posture but does not necessarily become manifest.
<i>Ocular motility</i>	The primary underaction of the affected lateral rectus results in limited abduction and is followed by a number of sequelae. There is overaction of both the contralateral and ipsilateral medial recti. There is also secondary underaction of the contralateral lateral rectus (Fig. 14.8).
<i>Binocular function</i>	This is often retained in the presence of an abnormal head posture. As the angle of deviation is often smaller for near fixation, binocular function is usually present on near fixation. In cases of head trauma, fusion may have been lost.
<i>Hess chart</i>	This provides a record for the notes and illustrates the associated muscle sequelae (Fig. 14.9).

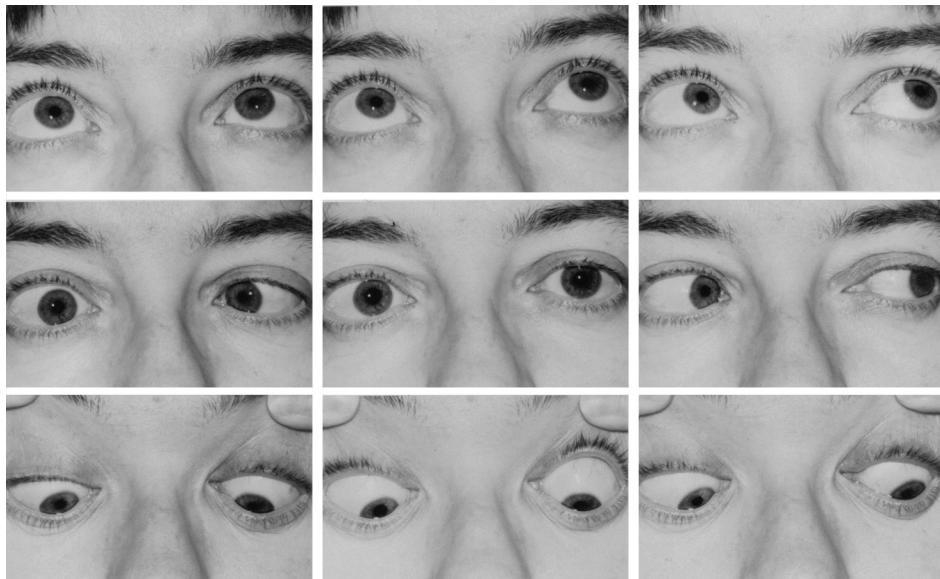


Figure 14.8 Right VI nerve palsy.

Field of binocular single vision

The field of binocular single vision is displaced away from the affected side (Fig. 14.10).

Diplopia

This is uncrossed and becomes worse for distance fixation.

Differential diagnosis

Differential diagnosis should be made with the following conditions: Duane's retraction syndrome, infantile esotropia, nystagmus block syndrome and medial wall fracture.

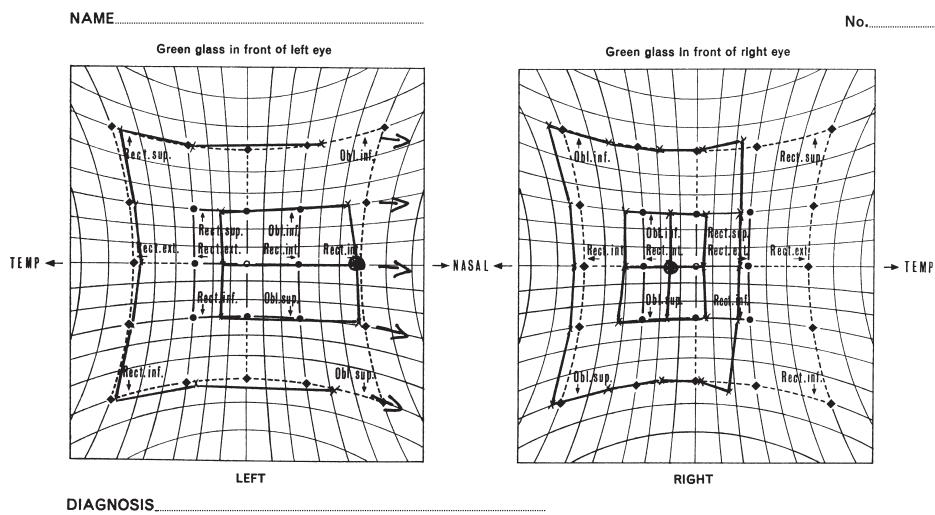


Figure 14.9 Hess chart of right VI nerve palsy.

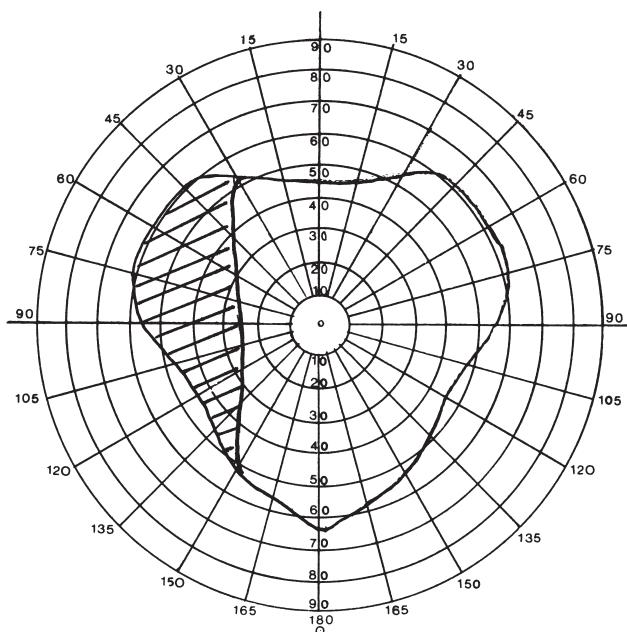


Figure 14.10 Field of binocular single vision of right VI nerve palsy.

Management

Recovery of VI nerve palsy can be expected in at least 50% of cases although a higher percentage of recovery is likely in those cases due to vascular insults, trauma and inflammation (Rucker 1966, Rush & Younge 1981). Good recovery of cranial nerve palsies following raised intracranial pressure is also expected. However, poor recovery is more likely with an aetiology of space-occupying lesions where the nerve is involved in the mass lesion with compression of nerve fibres or stretching of the nerve resulting in shearing injuries or contusion.

Congenital

Treat amblyopia if indicated. Surgery may be required for cosmesis (see below). In young children, treatment is aimed at preventing amblyopia and preserving binocular function (Rosenbaum 1991, Arochane & Repka 1995).

Children often suppress, leading to amblyopia. Therefore occlusion is required. If they adopt an abnormal head posture, this should not be discouraged. Development of concomitant strabismus may occur following acquired VI nerve palsy in children and related factors include poor visual acuity and visual field loss (Rowe & Noonan 2002).

Acquired

Treatment of the underlying cause of the VI nerve palsy if possible. In adult cases, allow for recovery but during this period, *prisms or occlusion* can be used to join

or eliminate diplopia. Where small-angle deviations are present, it may be possible to incorporate prisms into a spectacle correction once the deviation is static, to provide long-term management. Fresnel prisms are useful for small deviations. However, vision may be blurred by the prisms in proportion to the amount of prism applied.

Surgery may be employed once the condition is stable and static with a minimum of 6 months of stable measurements. Depending on the size of the angle of deviation, unilateral but more commonly bilateral medial recti recessions may be required (Gonzalez *et al.* 2005). The lateral rectus may be resected in addition (Rosenbaum *et al.* 1989).

If the palsy is complete, a medial rectus recession can be combined with muscle transposition of the superior and inferior recti to the affected lateral rectus (Berens & Girard 1950, Parks 1974, Metz 1976, Wybar 1981, Ciancia *et al.* 1984). Transposition surgery increases the size and improves the location of binocular single vision but does not achieve full ocular rotation: a reduction in abduction and adduction is to be expected (Murgatroyd *et al.* 2002). Three-muscle surgery increases the risk of anterior segment ischaemia and unaugmented full tendon vertical rectus transposition surgery without medial rectus recession can also be considered (Bansal *et al.* 2006).

Where there is residual lateral incomitance, a recession or faden on the contralateral medial rectus may be considered.

Botulinum toxin A may be injected into the medial rectus muscle in recent onset palsies. This helps prevent contracture and allows the eyes to adopt a straighter position in order to obtain binocular single vision. It may help to restore binocular function in the long term (Scott *et al.* 1989, Scott *et al.* 1990). The use of botulinum toxin A may be combined with surgery when performing a transposition procedure (Fitzsimons *et al.* 1988, 1989).

Prospective randomised clinical trials have shown no evidence of any difference between the treated and untreated groups in terms of ultimate outcome. However, botulinum toxin A treated patients reported subjective improvement (Lee *et al.* 1994).

Multiple sclerosis

Aetiology

Multiple sclerosis is an autoimmune disease in which there is chronic inflammatory demyelinating disease of the central nervous system (Bruck 2005). If there is inflammation, there are also biochemical changes. Impaired nerve conduction is a consequence to loss of myelin along the nerve sheath and formation of sclerotic plaques can be seen on neuroimaging of the brain. Improvement occurs because inflammation abates and there is a degree of remyelination and reorganisation of cell functions such as an increase in sodium channels. There is also plasticity of the nervous system with rerouting of impulses.

Investigation

The orthoptic investigation is dependent on the type of ocular motility defect that has occurred. Generally, there are isolated muscle palsies, internuclear ophthalmoplegia and nystagmus, saccadic, smooth pursuit and vestibulo-ocular impairments, gaze palsies and skew deviation (Morris & Rowe 2007). Motion perception difficulties (Pulfrich's phenomenon) may also be reported that may relate to a mismatch in the relay of visual information through a damaged visual pathway (Derrington 2000). Ophthalmic symptoms are reported in approximately 85% of cases and are present at the initial presentation in roughly 50% (Steinlin *et al.* 1995).

The prognosis for patients with normal baseline brain scans is very good with few developing multiple sclerosis. Most cases with abnormal scans at baseline assessment develop multiple sclerosis. The more lesions at baseline, the more severe and disabling the condition will be in the long term.

Patients develop recurrent attacks of multifocal neurological deficits that are typically of the spinal cord, optic nerve and brain. There are different types of multiple sclerosis that include relapsing remitting, primary progressive and secondary progressive (Armstrong 1999).

Management

For an isolated episode, the use of steroids settles the episode faster. These are given intravenously for 3 days if the patient has shown little recovery after 7–10 days. Interferon over 18–24 months at a low dose may delay conversion to the chronic disease.

Counselling should address issues such as the risk of developing multiple sclerosis, patient access to medical information and financial/insurance issues.

Relapse management includes the use of steroids, multidisciplinary involvement of nurses, occupational therapists, physiotherapists and the orthoptist. Symptoms must be actively managed while waiting for recovery and treatment of the ocular motility deficit is with prisms, occlusion and/or surgery as indicated. Blue lenses or neutral density filters may be considered for patients with motion perception difficulties (MacIntosh 2003).

Mitoxantrone may be effective for switching off aggressive relapsing multiple sclerosis but cannot be used long term due to its toxicity. However it stabilises, patients and further treatment can be instigated at that point.

Acquired motor fusion deficiency

Aetiology

This occurs after closed head trauma, after cerebrovascular accidents, as a result of intracranial tumours and after brain surgery (Avilla & von Noorden 1984). There is an assumed midbrain lesion. It may also occur after prolonged sensory

deprivation following cataract (Pratt-Johnson & Tillson 1988, Sharkey & Sellar 1994).

Investigation

<i>Case history</i>	Patients complain of severe asthenopia, intractable diplopia and inability to maintain single vision for any length of time. Diplopia may be crossed, then uncrossed, and then vertical.
<i>Cover test</i>	Involuntary vertical bobbing movements of the non-fixing eye may be seen with both eyes open. This occurs at or near the angle of superimposition (Pratt-Johnson & Tillson 1988).
<i>Accommodation</i>	This may be associated with a reduced range of accommodation (Wade 1965, Pratt-Johnson 1973, Pratt-Johnson & Tillson 1979, Harrison 1987).
<i>Binocular function</i>	There is no motor fusion. Sensory fusion and stereopsis are intermittently demonstrable (Avilla & von Noorden 1984).

Management

There is no effective treatment. Prisms are not useful due to variable diplopia and patients cannot use motor fusion once aligned. There is no suppression area to move the image in to.

The condition can spontaneously improve but this is uncommon (Hart 1969, MacLellan 1974, Lee 1998). Filters, occlusion or occlusive contact lens are required to alleviate the diplopia (McIntyre & Fells 1996). Occlusion may also be provided in the form of an occlusive intraocular lens or by unilateral aphakia (Pratt-Johnson & Tillson 1988).

Non-accidental injury

Shaken baby syndrome or non-accidental injury has a high risk of mortality (Kilvin *et al.* 2000). Associated signs and symptoms are multiple and require careful ophthalmological and neurological examination. Visual impairment results from cortical and ocular damage (Rahi & Cable 2003). In addition, there is reduced functional mobility, behavioural issues and long-term neurological sequelae.

Associated signs and symptoms include the following (Coody *et al.* 1994, McCabe & Donahue 2000):

- Bradycardia
- Diarrhoea
- Hypothermia
- Irritability
- Hypotonia
- Seizures

- Bulging fontanelle
- Reduced levels of consciousness
- Hydrocephalus
- Subdural haematoma and apnoea

Classic ocular signs include the following (Ludwig & Warman 1984, Green *et al.* 1996, Kilvin *et al.* 2000, McCabe & Donahue 2000, Morad *et al.* 2002, Mungan 2007):

- Retinal haemorrhages
- Retinoschisis
- Retinal folds
- Vitreous traction
- Vitreous haemorrhage
- Optic nerve sheath haemorrhage
- Subluxed or dislocation lens
- Glaucoma
- Cataract
- Anterior chamber regression
- Iridodialysis
- Retinal detachment
- Poor visual acuity
- Papilloedema
- Anisocoria
- Gaze preference
- Nystagmus
- Cranial nerve palsy

Premature visual impairment

Increasing survival of babies born prematurely has resulted in an increase in children with long-term disabilities (Johnson 2007). Extremely low birth weight babies are those with a birth weight of <1001 g. Low birth weight babies have a weight of >1501 g but <2500 g. Preterm babies are at risk of developing abnormalities of physical, cognitive and behavioural functions (Wong *et al.* 2008). Visual impairment may be caused by neurological damage but also by retinopathy of prematurity.

Investigation

Refraction

There is an increased prevalence of refractive errors in these babies with failure to emmetropise (O'Connor *et al.* 2002, O'Connor *et al.* 2006).

Fundus and media

Often, features of retinopathy of prematurity are present.

<i>Visual acuity</i>	Uniocular and binocular visual acuity is reduced at near and distance fixation compared to full-term babies. Impaired colour vision, visual field and contrast sensitivity may be noted (Dobson <i>et al.</i> 1996, Larsson <i>et al.</i> 2006, O'Connor & Fielder 2007).
<i>Cover test</i>	Strabismus is common.
<i>Ocular motility</i>	Fixation and smooth pursuit abnormalities may be noted (Cioni <i>et al.</i> 1997).

Ophthalmoplegia

This is a group of conditions that have a variety of causative factors, where there is a paresis of two or more extraocular muscles (Lueck 2011). Ophthalmoplegia is not only present in third nerve palsy but also where there is combined cranial nerve palsies such as fourth and sixth. Where more than one cranial nerve is involved, this suggests that pathology relates to an anatomical area where the cranial nerves are in close proximity, such as the cavernous sinus, orbital apex and superior orbital fissure.

Cavernous sinus syndrome

There is a lesion in the cavernous sinus causing paresis of the ocular motor nerves (III, IV and VI) and the first two divisions of the fifth nerve (Hunt *et al.* 1961).

Aetiology

- Tumour affecting both cavernous sinuses (Barry & Schroeder 2010)
- Cavernous sinus thrombosis
- Cavernous sinus fistula
- Cavernous sinus inflammation

Investigation

<i>Case history</i>	Diplopia is due to ophthalmoplegia and reduced visual acuity. There is often a marked conjunctival injection. An ocular bruit may be present, which is audible with the use of a stethoscope.
<i>Cover test</i>	Proptosis is often present that may be pulsatile. Strabismus relates to the cranial nerves involved.
<i>Intraocular pressure</i>	Raised intraocular pressure is related to the proptosis.
<i>Ocular motility</i>	Pupillary reaction is often spared. Ophthalmoplegia is due to involvement of the III, IV and VI nerves. Ptosis may be evident.
<i>Sensory function</i>	Involvement of Va and Vb can lead to impaired sensation on either side of the face and reduced corneal sensitivity.

Sphenoidal fissure syndrome

Aetiology

A lesion in the region of the superior orbital fissure may affect any structure passing through.

Investigation

<i>Case history</i>	Diplopia is due to III, IV and VI cranial nerve involvement. Symptoms are variable depending on the extent and combination of nerve involvement. Pain and anaesthesia is due to involvement of the V cranial nerve.
<i>Visual acuity</i>	This may be reduced if there is optic nerve compression.
<i>Ocular motility</i>	There is progressive involvement of the III (sometimes only the superior branch), IV and VI cranial nerves. Proptosis is noted.
<i>Sensory function</i>	Involvement of Va and Vb can lead to impaired sensation on either side of the face and reduced corneal sensitivity.

Orbital apex syndrome

Aetiology

A lesion in the posterior part of the orbit near its apex is responsible for this condition. Orbital pseudo tumour may produce this although visual loss is infrequent. Orbital cellulitis may also produce this syndrome.

Investigation

<i>Case history</i>	V nerve involvement will give symptoms of severe pain. Diplopia is due to defective ocular motility.
<i>Visual acuity</i>	Reduced visual acuity is due to compression of the optic nerve.
<i>Ocular motility</i>	Limited ocular movement is seen with III, IV and VI nerve involvement according to the muscles involved. There is often marked swelling and redness of the eyelids with chemosis of the conjunctiva. Proptosis and raised intraocular pressure are also documented.

Guillain–Barre syndrome – Fisher's syndrome

Aetiology

This syndrome is thought to be the result of an allergic response of the peripheral nervous system to an antigen, possibly of viral origin. There is acute demyelinating

polyneuropathy with failure of the immune system and with potential life-threatening consequences (Güngör *et al.* 2011, Hardy *et al.* 2011). Fisher's syndrome is a variant with less severe clinical features.

Investigation

<i>Ocular motility</i>	Facial nerve paralysis may be noted. There is mild to complete ophthalmoplegia and ptosis, anisocoria and nystagmus may be noted.
<i>Visual acuity</i>	Optic neuritis may occur with papilloedema evident on fundus examination.
<i>Associated paralysis</i>	There may be variable paralysis of motor nerves beginning in the lower extremities with absent tendon reflexes. There may also be involvement of the respiratory muscles requiring ventilation in intensive care units during the acute phase.

Recovery is a slow and gradual process, but in general the prognosis is favourable. Fisher's syndrome is characterised by total external ophthalmoplegia, gait ataxia and hyporeflexia. Ophthalmoplegia involves symmetrical impairment of conjugate upward and lateral gaze progressing frequently to complete ophthalmoplegia. There may be facial nerve involvement and rarely internal ophthalmoplegia occurs with blurred vision and bilateral tonic pupils (Bae *et al.* 2009). There is absence of limb weakness and patients usually recover spontaneously within 7–12 weeks. Treatment is with intravenous immunoglobulin. The condition can recur (Orr & Storey 2004).

Ophthalmoplegia migraine

This condition involves migraine and is usually an associated III nerve palsy without pupil sparing. Other oculomotor nerves may also be affected leading to corresponding muscle paresis (Horne *et al.* 2002).

Aetiology

It is postulated that the III nerve is compressed in the cavernous sinus by a dilated or oedematous carotid artery but this is not a consistent finding. Alternatively, vasospasm may affect the small vessels supplying the nerve itself.

Investigation

A typical episode consists of pain in and around the involved eye. There may be associated nausea and vomiting. With the onset of ophthalmoplegia, the head pain often resolves. The paresis usually completely resolves within 1 month but there may be permanent full or partial disability. The onset is usually in childhood and there is often a positive family history of migraine.

Tolosa–Hunt syndrome

This is an acute inflammatory condition that involves the superior orbital fissure or the anterior cavernous sinus resulting in a painful ophthalmoplegia.

Aetiology

There may be non-specific granulation inflammation in the cavernous sinus or pachymeningitis of the superior orbital fissure due to an inflammatory or allergic inflammation process (Aron-Rosa *et al.* 1978).

Investigation

<i>Case history</i>	The patient may complain of a steady, boring pain in or around the eye. However, some cases are painless (Cornish <i>et al.</i> 2011). Diplopia may be appreciated. Hypoaesthesia or anaesthesia in the distribution of the ophthalmic division of the V nerve may be experienced.
<i>Visual acuity</i>	Occasionally, there is a reduction in visual acuity.
<i>Ocular motility</i>	Ophthalmoplegia with complete or partial palsy of extraocular muscles may be documented and the pupil may or may not be affected. The limited ocular movements can be bilateral (Sugie <i>et al.</i> 2011).
<i>Sensory function</i>	Involvement of Va can lead to impaired sensation on either side of the face and reduced corneal sensitivity (Navi & Safdieh 2010).

The condition occurs acutely or sub-acutely and usually responds to high doses of corticosteroids. It is relapsing and remitting and spontaneous remission may occur with complete or partial recovery (Navi & Safdieh 2010).

Ocular neuromyotonia

This is a rare condition resulting in transient, aperiodic ocular misalignment.

Aetiology

There is spontaneous firing of ocular motor nerves resulting in impairment of muscle relaxation. The spontaneous firing is thought to occur in injured axons or with radiation plexopathy and peripheral trauma neuropathy. Many patients have had prior radiation therapy to the sellar and parasellar region or as a consequence to vascular abnormalities (Fricke *et al.* 2002). The condition is particularly noted following pituitary fossa surgery and irradiation or after external beam radiotherapy and radiosurgery (Much *et al.* 2009). Experimental evidence has implicated extracellular potassium concentration in causing the spontaneous firing of axons.

Investigation

<i>Case history</i>	There is episodic diplopia, which occurs either spontaneously or after sustained eccentric gaze. It can be the presenting sign of an intracranial tumour (Salchow & Wermund 2011).
<i>Ocular motility</i>	There is often a cranial nerve palsy and a third nerve palsy is most commonly noted with aberrant regeneration (Choi <i>et al.</i> 2006). Abducens neuromyotonia is also reported (Kau & Tsai 2010, Salchow & Wermund 2011). Attacks can be provoked by overaction of the muscles supplied by the cranial nerve involved and therefore may be elicited by sustained gaze in that direction of gaze (Spiritus <i>et al.</i> 1995, Ezra <i>et al.</i> 1996).
<i>Differential diagnosis</i>	A differential must be made for myasthenia gravis, convergence spasm and cyclic third nerve palsy.
<i>Electrophysiology</i>	The diagnosis is supported by electrophysiological studies (Salchow & Wermund 2011).

Management

Treatment with anti-convulsants is extremely effective. Carbamazepine is most frequently employed.

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15 Mechanical Paralytic Strabismus

Mechanical strabismus is paralytic strabismus, which is caused by factors that interfere with muscle contraction or relaxation, or prevent free movement of the globe.

There may be a physical restraint that interferes with normal function and is sometimes referred to as a leash. This may be of the following types:

- *Direct*: For example, tight muscles. This limits movement in the position of gaze opposite to the leash.
- *Indirect*: For example, posteriorly inserted muscle. This limits movement in the position towards the leash.

Classification

Congenital

- Congenital cranial dysinnervation disorders:
 - Duane's retraction syndrome
 - Congenital fibrosis of the extraocular muscles
 - Synergistic divergence
- Brown's syndrome
- Adherence syndrome
- Moebius syndrome
- Strabismus fixus syndrome

Acquired

- Brown's syndrome
 - Adherence syndrome
 - Thyroid eye disease
 - Orbital injuries
 - Orbital inflammation
- } causing muscle fibrosis and development
} of fibrotic tethers

- Conjunctival shortening syndrome
- Retinal detachment
- Macular translocation surgery
- Cataract
- Space-occupying lesions
- Iatrogenic

Characteristics

1. The deviation in the primary position and the abnormal head posture are generally small.
2. Ocular motility:
 - a. Ductions and versions are equally limited.
 - b. Movement in the opposite direction of gaze is often limited.
 - c. Muscle sequelae are often limited to overaction of the contralateral synergist.
 - d. Abnormal patterns of movement may be present such as upshoots.
 - e. Normal saccadic velocity is present up to the point of limitation.
 - f. Pain on movement may be experienced.
3. Forced duction test shows limitation of passive movement.
4. Globe retraction is diagnostic of mechanical restrictions.

Management

The primary aim of management is to ensure that the patient is symptom-free and the deviation is cosmetically satisfactory.

Treatment is indicated where the condition produces symptoms and there is poor cosmesis.

Aims of surgical treatment

- To centralise and enlarge the field of binocular single vision and therefore reduce the abnormal head posture.
- Obtain good cosmesis.
- Reduce corneal exposure due to absence of Bell's phenomenon.
- Reduce or eliminate any obvious abnormal pattern of movement.

Principles

- Reduction of the mechanical restriction.
- Weaken the overacting contralateral synergist to induce relative concomitance.
- Decrease abnormal movement by stabilising the positions of other muscles.

Congenital cranial dysinnervation disorders

Duane's retraction syndrome

This is a congenital condition although it has been described as an acquired condition in rare cases. The condition may be bilateral (Rowe *et al.* 1991, Zanin *et al.* 2010) or unilateral, the left eye is more commonly affected, and the incidence is higher in females. It occurs in 1–5% of strabismic patients.

Classification

Brown (1950)

The condition is classified according to the clinical findings:

- *Type A*: Typical convergent deviation with limited abduction and limited adduction to a lesser degree
- *Type B*: Typical convergent deviation with limited abduction and normal adduction
- *Type C*: Atypical divergent deviation with limited adduction and limited abduction to a lesser degree

Huber (1974)

The condition is classified on the basis of electromyography recordings and the co-contraction theory where the lateral rectus receives co-innervation from the III and VI nerves:

- *Type I*: There is limited abduction and limited adduction to a lesser degree. The lateral rectus has little or no innervation on abduction but has some innervation on adduction.
- *Type II*: There is limited adduction. The lateral rectus has innervation on abduction and adduction. Therefore, it can abduct, and on adduction it counteracts the medial rectus producing limited adduction and globe retraction.
- *Type III*: There is limited abduction and adduction. The lateral rectus has little or no innervation to abduct, but is innervated on adduction and therefore counteracts the medial rectus.

Aetiology

Mechanical

- Thin, elastic muscles
- Muscles bound to the orbital wall
- Posterior insertion of the medial rectus

- Abnormal attachments from the medial rectus to the orbital apex resulting in a tight medial rectus unable to relax or contract
- Inelastic, fibrotic lateral rectus with abnormal attachments

Innervation

- *Absent VI nucleus:* The lateral rectus shows atrophy and fibrosis (Hotchkiss *et al.* 1980, Parsa *et al.* 1998).
- *Partially formed VI nerve.*
- *Co-contraction innervation theory:* Magnetic resonance imaging shows that the lateral rectus is partially innervated by the III nerve with or without VI nerve innervation and with additional hypoplasia of the superior oblique, superior rectus and levator palpebrae superioris muscles in some cases (Demer *et al.* 2007, Yanghong *et al.* 2009). This produces co-contraction of the medial and lateral recti on adduction and limited adduction with retraction of the globe.

Investigation

<i>Case history</i>	Children present because parents have noted abnormal eye movements, head posture and/or strabismus. Adults may present with symptoms of decompensation. Diplopia is rarely noticed.
<i>Visual acuity</i>	Amblyopia may be present if there is associated manifest strabismus or anisometropia.
<i>Abnormal head posture</i>	A face turn to the affected side is most often seen.
<i>Cover test</i>	This is performed with and without the abnormal head posture. A typical condition has a convergent deviation and the atypical condition has a divergent deviation.
<i>Ocular motility</i>	Limited abduction occurs to a variable degree and limited adduction occurs often to a lesser degree. Retraction of the globe on adduction with narrowing of the palpebral fissure is often noted. Widening of the palpebral fissure may be seen on attempted abduction. Upshoots or downshoots of the affected eye may occur on adduction with depression or elevation of the affected eye on abduction and bulging of orbital fat through the orbital septum may occur as the eye is adducted. An A or V pattern may be documented. Vertical retraction syndrome is a rare variant where there is limited elevation with lid retraction and limited depression with narrowing of the palpebral fissure (Fig. 15.1).
	Upshoots are more common than downshoots and occur because of globe slippage between the tight horizontal recti (bridal effect) or because of slippage of the tight horizontal recti over the globe.



Figure 15.1 Duane's retraction syndrome.

*Convergence
Binocular
function*

*Angle of
deviation
Hess chart
Field of
binocular
single vision
Differential
diagnosis*

*Associated eye
signs*

Bilateral syndromes have a higher incidence of A or V patterns with horizontal and vertical ocular motility deficits (Zanin *et al.* 2010).

This is poor if there is limited adduction.

There is usually a good level of binocular function present, which is maintained with an abnormal head posture. A mixture of binocular and suppression responses is recorded on clinical testing.

Primary and secondary angles differ indicating incomitance.

Retention of incomitance is documented (Fig. 15.2).

The area of binocular single vision is positioned away from the position of maximum limitation (Fig. 15.3).

It is necessary to ensure that the deviation is not due to a VI nerve palsy, medial wall fracture, localised inflammation, Moebius syndrome or iatrogenic cause such as a liberal lateral rectus recession.

Associated eye signs include lens opacities, heterochromia irides, fundal defects, microphthalmos and crocodile tears (Ramsay & Taylor 1980).

NAME

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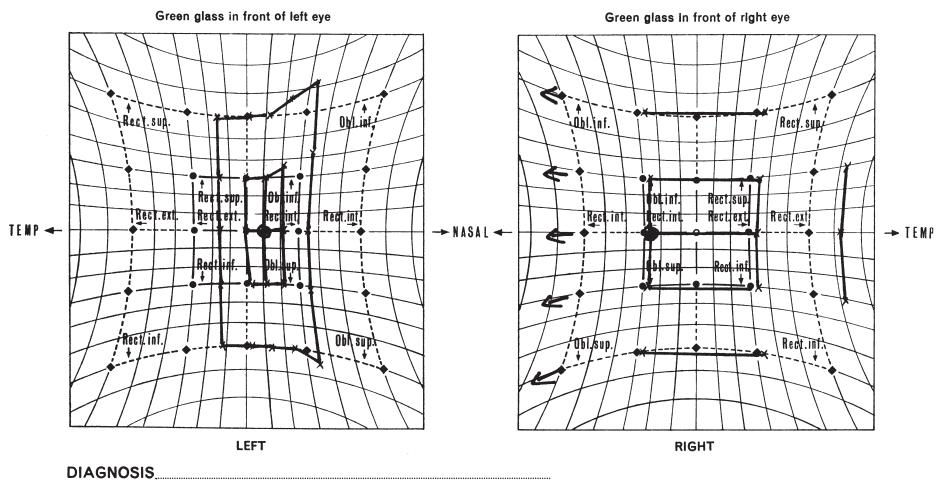


Figure 15.2 Hess chart of Duane's retraction syndrome.

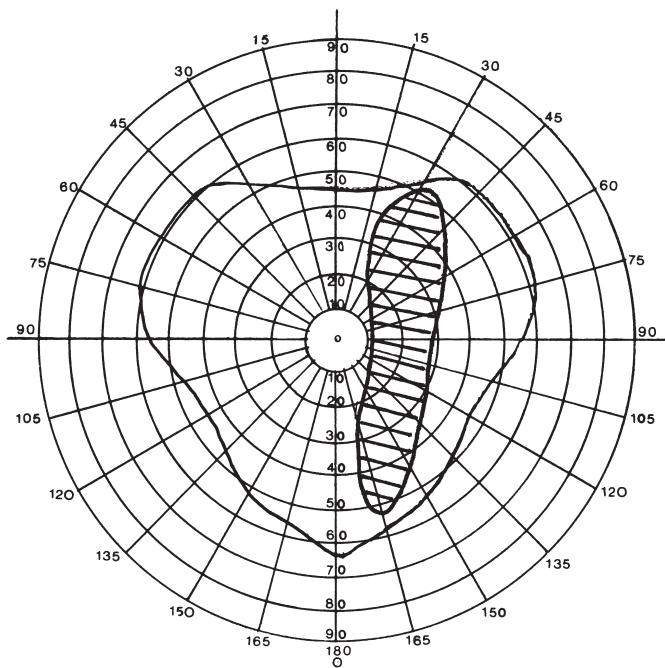


Figure 15.3 Field of binocular single vision of Duane's retraction syndrome.

Associated general signs	<p>Two associated general conditions are those of Klippel–Feil and Goldenhar (Velez 1970).</p> <ul style="list-style-type: none">• <i>Goldenhar's:</i> The patient has ocular, auditory and vertebral anomalies with a poor mental performance (Pieroni 1969). Signs include hypoplasia of the face, low set ears, deafness, coloboma, short neck, midline facial cleft and dermoid cysts (Caca <i>et al.</i> 2006).• <i>Klippel–Feil:</i> The patient has a short, immobile rigid neck with a low posterior hairline.• <i>Wildervanck syndrome:</i> This syndrome comprises Duane's retraction syndrome, Klippel–Feil and deafness with cleft palate and short stature (Kumar <i>et al.</i> 2010).• <i>Kirkham's triad:</i> The rare triad is composed of Duane's retraction syndrome, Klippel–Feil and perceptive deafness. This is inherited as a single gene defect (Kirkham 1969). There is congenital fusion of any two of the seven cervical vertebrae. The syndrome is caused by failure in normal segmentation or division of cervical vertebrae during the early weeks of foetal development. Signs include a short neck, low hairline at the back of the head and restricted mobility of the upper spine.
DURS2	<p>This is a dominant Duane's retraction syndrome in which there is bilateral type I and/or type III features. An A or V pattern may be seen. There is always sixth nerve hypoplasia but there may also be superior oblique and superior rectus muscle hypoplasia, optic nerve hypoplasia and third nerve hypoplasia (Demer <i>et al.</i> 2007).</p>

Management

Correct the refractive error if present and treat any amblyopia. There is usually no further treatment as the majority of patients are comfortable and remain compensated, often with only a slight abnormal head posture (Chua *et al.* 2005). Symptoms are uncommon.

Indications for treatment

- Decompensation resulting in strabismus in children and symptoms in adults.
- Cosmetically poor abnormal head posture.
- Cosmetically poor strabismus.

Aims of treatment

- Place the field of binocular single vision more centrally and enlarge the field where possible.
- Reduce or eliminate the abnormal head posture.
- Achieve a good cosmesis.

Prisms

Prisms may be used first in cases of decompensation to assess if correction of the deviation helps or if the abnormal head posture is helped. They are given as an alternative method of treatment if possible.

Botulinum toxin

This can be used diagnostically and therapeutically. For diagnostic purposes, botulinum toxin can be used to determine whether reduction of angle of deviation will improve binocular function, reduce abnormal head posture or reduce diplopia (Dawson *et al.* 2010). Some patients choose maintenance botulinum toxin rather than surgery as a long-term treatment option.

Surgery

The purpose of surgery is not to obtain an improvement of ocular movement and it is unlikely that any significant increase in abduction or adduction will occur post-operatively.

The forced duction test will give a positive result due to the presence of fibrotic muscles. It is important to never resect the lateral rectus in either eye as this would result in further restriction of ocular motility.

Typical (eso-deviation)

Ipsilateral or bilateral medial rectus recessions are advocated depending on the degree of the deviation. For large-angle eso-deviations and retraction, bilateral medial recti and ipsilateral lateral rectus recessions are useful.

Fells (1971) has used temporal transfer of superior and inferior recti to the lateral rectus combined with ipsilateral medial rectus recession <3.5 mm. This procedure increases abduction and reduces eso-deviation and abnormal head posture (Rosenbaum 2004, Sterk *et al.* 2004, Britt *et al.* 2005).

Atypical (exo-deviation)

Ipsilateral or bilateral lateral rectus recessions are performed depending on the degree of the deviation. Vertical transposition surgery of the superior and inferior rectus muscles to the medial rectus may be undertaken to improve adduction (Snir *et al.* 2005).

Upshoots/downshoots and globe retraction

In the Faden procedure globe slippage is prevented by adding fixation suture to the medial rectus and lateral rectus. This prevents the globe from slipping as the globe is tethered to the horizontal muscles.

Globe retraction and upshoots or downshoots are dealt with effectively with ipsilateral medial and lateral recti recessions (von Noorden 1992).

Upshoots and downshoots may also be innervational in origin and may be reduced with superior and inferior rectus recessions (Mohan & Saroha 2002).

Vertical retraction syndrome (Malbran 1963)

This is a rare syndrome where there is absence or impairment of elevation with lid retraction. To a lesser degree, there may be limitation of depression with narrowing of the palpebral fissure. This may be due to inferior rectus muscle tethering or midbrain pathology (Man *et al.* 2009).

Congenital fibrosis of the extraocular muscles (CFEOM)

This is a hereditary form of musculofascial anomaly, involving most or all of the extraocular muscles. The condition is present from birth, is non-progressive and primarily due to neurogenic disturbances of the brainstem or cranial nerve development (Sugawara *et al.* 1982). A total of seven disease genes and ten phenotypes have been identified with more awaiting gene elucidation (Oystreck *et al.* 2011).

Aetiology

The condition is due to fibrotic or degenerative changes in the extraocular muscles. There is autosomal dominant inheritance, which is of three types (Heidary *et al.* 2008):

1. *CFEOM1*: Typical type with dominant inheritance (maps to chromosome 12). Features include bilateral ptosis, limited elevation and strabismus.
2. *CFEOM2*: Recessive inheritance with congenital, bilateral ophthalmoplegia and ptosis and exotropia (Bosley *et al.* 2006).
3. *CFEOM3*: Atypical type with dominant or recessive inheritance (multiple chromosome mutations). May have features of CFEOM types 1 and 2 but may have features including absent or unilateral ptosis, unilateral ophthalmoplegia and some elevation function (Demer *et al.* 2010).

Investigation

<i>Case history</i>	There is a positive family history. Signs are present from birth and are almost always bilateral.
<i>Visual acuity</i>	Amblyopia is frequent. Reduced vision may also be due to exposure keratitis following surgery for ptosis.
<i>Cover test</i>	CFEOM1 commonly has an A exotropia. The eyes are fixed in a depressed position with no elevation beyond the midline.
<i>Abnormal head posture</i>	Chin elevation is adopted.

<i>Ocular motility</i>	Severely limited movements are demonstrated, particularly on upgaze as the inferior muscles are affected first. There is often a spasm of convergence on elevation producing a shallow A pattern. The condition may be unilateral but is more commonly bilateral. Bilateral ptosis is symmetrical or asymmetrical and Bell's phenomenon is absent.
<i>Forced duction test</i>	The patient may have some nystagmus on attempted elevation. Positive due to extraocular muscle restrictions.

Management

Treat amblyopia where indicated. Surgery may be required for strabismus or ptosis. The aim is to remove obstacles to the visual axes in primary gaze, alleviate abnormal head posture and align the eyes in primary gaze. A forced duction test is required pre-operatively and during surgery. Surgery may involve extraocular muscle extirpation for those that lack innervated antagonist muscles.

However, strabismic surgery tends to have disappointing results. The procedure usually involves an inferior rectus recession and/or superior rectus resection and the use of stay sutures may be considered. Large recessions and hang back sutures are used as necessary with careful extensive posterior dissection of the rectus muscles from the intermuscular septal attachments. Conjunctival recession may also be undertaken.

Ptosis surgery, if undertaken, should only partially correct the ptosis as where there is an absent Bell's phenomenon, full correction of the ptosis may result in corneal exposure.

A conservative fascia lata frontalis sling procedure is advised and surgical results for ptosis are generally good (Magli *et al.* 2003).

Synergistic divergence

This is a distinct but rare pattern of ocular motility in which both eyes have severely limited adduction with large-angle exotropia. Divergence of the eye increases on contralateral gaze. It is proposed that this is due to denervation of the medial rectus muscle coupled with abnormal innervation of the ipsilateral lateral rectus muscle by the third cranial nerve (Oystreck *et al.* 2009). Individuals may have an absent VI nerve and hypoplastic III nerve (Kim & Hwang 2009).

Brown's syndrome

The natural history of Brown's syndrome shows that there is clear evidence for spontaneous improvement in some cases (Adler 1959, Dyer 1970, Scott & Knapp

1972). The syndrome may be unilateral or bilateral. There is some evidence of genetic inheritance for Brown's syndrome and in association with other ocular motility disorders (Rowe & Noonan 2006).

Aetiology

Congenital

1. Short anterior superior oblique tendon sheath.
2. Tight, inelastic superior oblique tendon. Sometimes the musculofacial anomaly is posterior to the globe and is not found on surgical exploration.
3. A swelling or nodule on the superior oblique tendon will prevent free movement of the muscle through the trochlea. The click syndrome occurs because the nodule is forced through the trochlea. Such a swelling may be found on surgical exploration of the trochlear area (Maggi & Maggi 2002).
4. Anomalous innervation to the superior and inferior oblique muscles of the affected eye.
5. Bifid sclera insertion of the superior oblique muscle (Park *et al.* 2009).

Acquired

1. Injury to the trochlea leading to scar tissue formation. This may be from a penetrating injury to the trochlea area or a fracture in this area (Baker & Conklin 1987, Seo *et al.* 2010).
2. Rheumatoid conditions that have developed in later life (Killian *et al.* 1977).
3. Swelling on tendon.
4. Inflammation spreading from ethmoid (Hermann 1978).
5. Iatrogenic following superior oblique placation surgery (Helveston & Ellis 1983) or following frontal sinus surgery (Blanchard & Young 1984).

Investigation

Case history Children present as parents notice abnormal eye movement, head posture and strabismus. Adults present as they appreciate diplopia, pain, discomfort, tugging around trochlea area, palpable or audible clicks. Associated conditions may include rheumatism or trigger-finger syndrome. There may be a history of inflammatory conditions, trauma or previous surgery (involving a superior oblique tuck procedure) (Roper-Hall & Roper-Hall 1972).

Visual acuity Amblyopia may be present if manifest strabismus is present (Wright 1999).

Abnormal head posture The patient may have a slight chin elevation, head tilt to the affected side and face turn away from the affected side. The head posture is rarely marked.

Cover test

A small primary position hypo-deviation of the affected eye is demonstrated, which is commonly latent. Acquired Brown's syndrome may have significantly large hypotropia in primary position.

Ocular motility

There is limitation of the affected eye on elevation in adduction and often the eye cannot elevate beyond the midline. There is generally slight limitation on direct elevation. Limited elevation in abduction may be seen where the superior oblique is very tight (Wright 2009). A downshoot of the affected eye occurs on adduction. Muscle sequelae are limited to overaction of the contralateral synergist. A V pattern phenomenon is likely where the eyes diverge on elevation due to the abducting action of the inferior oblique muscle as it attempts to elevate the eye. Occasionally, an A pattern may be noted. Sometimes, the movement of the affected eye increases on repeated testing and may show improvement of the defective elevation. There may be a click and the eye may remain in an elevated position if the muscle has managed to pass through the trochlea (Fig. 15.4).



Figure 15.4 Right Brown's syndrome.

<i>Binocular function</i>	Binocular single vision is usually present in the primary position and in the lower field.
<i>Hess chart</i>	Retention of incomitance is documented. This shows inferior oblique underaction and overaction of the contralateral superior rectus only (Fig. 15.5).
<i>Field of binocular single vision</i>	Diplopia is limited to elevated positions of gaze (Fig. 15.6).
<i>Forced duction test</i>	Positive.
<i>Differential diagnosis</i>	

Investigation	Inferior oblique underaction	Brown's syndrome
<i>Abnormal head posture</i>	Marked head tilt	Slight head posture
<i>Angle in primary position</i>	Moderate	Slight
<i>Muscle sequelae</i>	Develops full sequelae	Limited to overaction of contralateral synergist
<i>A/V pattern</i>	A pattern	V pattern
<i>Forced duction test</i>	Negative	Positive

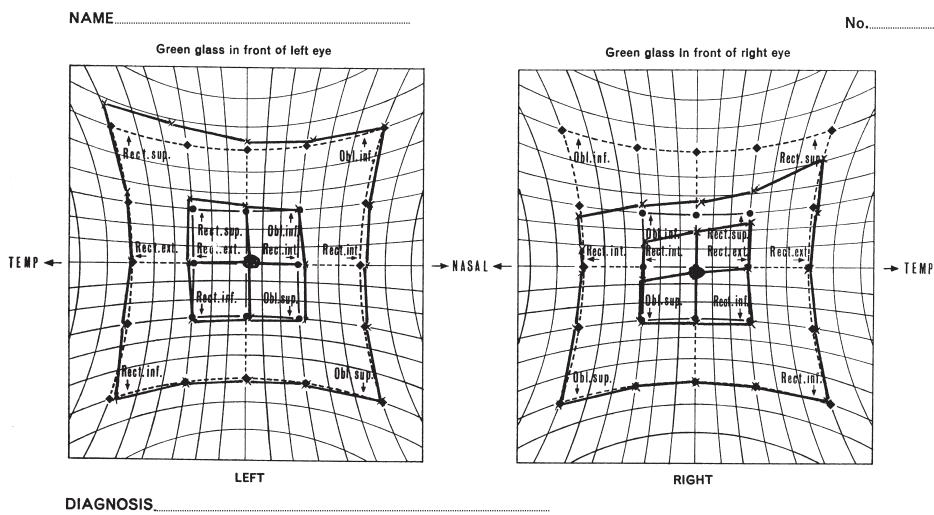


Figure 15.5 Hess chart of right Brown's syndrome.

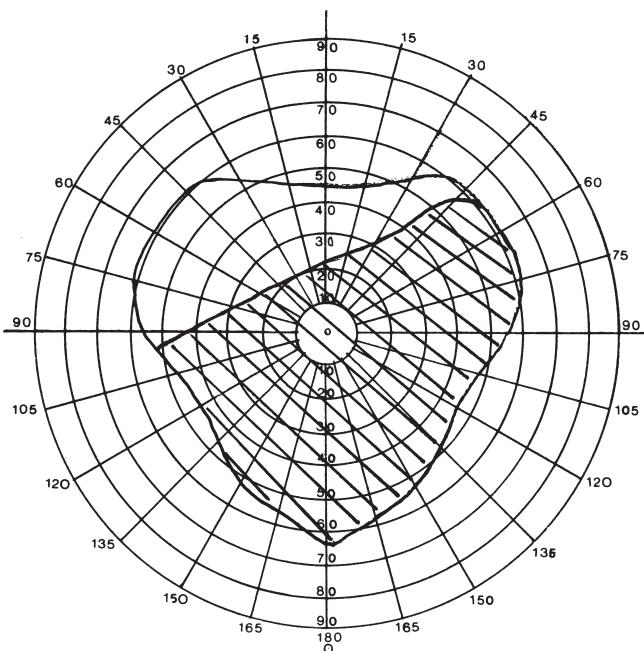


Figure 15.6 Field of binocular single vision of right Brown's syndrome.

Management

It is advisable not to intervene with invasive treatment unless there is noticeable chin elevation or vertical squint in primary position.

Natural history

Spontaneous improvement begins around 7 years of age and many cases show virtually full improvement by 12–15 years. The ‘click’ syndrome may be a phase in this natural improvement. Late spontaneous resolution may also occur in adulthood (Lambert 2010).

Correct the refractive error if present and treat amblyopia where indicated:

- **Local steroids:** They may be useful in acquired cases with inflammation (Saunders *et al.* 1990). These may be oral or injected corticosteroids (Hermann 1978, Beck & Hickling 1980).
- **Prisms:** They compensate for the deviation in the primary position but may be used with small-angle deviations only.

Indications for surgery

1. Diplopia in the primary position
2. Marked abnormal head posture

3. Decompensating deviation
4. Allows development of binocular single vision

Contraindications

1. Spontaneous recovery may occur
2. Presence of normal comfortable binocularity in the primary position
3. Only a slight abnormal head posture present

Surgery is rarely indicated and tends to be a last resort (Adler 1959). Surgery rarely improves the ocular rotations significantly (Sprunger *et al.* 1991).

The choice of surgery is governed by the result of the forced duction test and the findings at operation, which may establish the aetiology.

Procedures

1. Division and stripping of the superior oblique tendon sheath. This has been found to be insufficient on its own.
2. Superior oblique tendon elongation by intrasheath tenotomy, superior oblique posterior tenectomy, silicon tendon expanders or superior oblique tendon suture space (Stager *et al.* 1999, Stolovitch *et al.* 2002, Velez *et al.* 2006, Keskinbora 2007, Suh *et al.* 2008, Wright 2009).
3. Free any adhesions around the trochlear area. This may be combined with a complete superior oblique tenectomy. If a superior oblique paresis occurs, a contralateral inferior rectus recession may be performed as a second procedure.
4. Contralateral superior rectus recession.

Surgery does not usually produce immediate improvement in motility and can cause additional fibrosis and as a result, results may be disappointing.

Adherence syndrome

This syndrome results from a fusion of the fascial sheaths giving rise to various adherence syndromes.

Aetiology

Congenital

The syndrome is due to defective cleavage of the mesoderm where extraocular muscles are not differentiated from each other.

Acquired

The condition is due to trauma, inflammation or surgery. Where the cause is iatrogenic, there may be adherence of the muscle belly and globe, the muscle and

conjunctiva, or sclera and conjunctiva. This results from excessive bleeding, excessive cautery, multiple procedures, excessive tissue reaction, suture reaction, operative or post-operative infection.

Investigation

Ocular motility The type of defective ocular motility depends on the muscles or tendons that have adhered to each other (Johnson 1950).

The lateral rectus and inferior oblique adhering where they cross produce a pseudolateral rectus palsy.

Superior rectus and superior oblique adherence may produce a pseudosuperior rectus palsy.

Superior rectus and superior oblique adhering near the superior oblique insertion produce a pseudo superior oblique palsy with torsion (Kushner 2007).

Adherence of superior oblique tendon sheath and superior rectus produces a pseudo Brown's syndrome.

Adherence syndrome of Parks occurs after weakening of the inferior oblique, most frequently after myectomy. In this situation, the inferior oblique adheres to Tenon's capsule and produces hypotropia of the involved eye.

Adherence of the inferior oblique and inferior rectus muscles may occur following surgery on the inferior rectus, inadvertent inferior oblique tuck or from sclera buckling surgery (Kushner 2007).

Management

Management usually requires surgery where there is division of adhesions while the muscles are temporarily detached from the globe. Use of supramidextra implants in sleeve or sheet form after division of adhesions may prevent redevelopment of adhesions. Surgery may involve extensive conjunctival peritomy over 180°, recessing the conjunctiva while leaving the sclera to re-epithelialise (Burton *et al.* 2004). It is usually not possible to restore normal eye movements. Botulinum toxin may be used to aid alignment of vertical deviations (Barry *et al.* 2011).

Moebius syndrome

Aetiology

The congenital condition may be due to a primary developmental defect of the central nervous system with aphasia of the motor nuclei of the VI and VII nerves and denervation atrophy of facial and extraocular muscles. This is often associated with infranuclear defects (Lengyel *et al.* 2000, Cronemberger *et al.* 2001). There may be a primary mesodermal defect causing abnormal musculature with a

secondary supranuclear brainstem disorder. Brainstem necrosis occurs due to vascular deficiency (Spierer & Barak 2000).

Investigation

<i>Case history</i>	There may be an association with Poland syndrome and limb abnormalities (Miller & Strömland 1999). Speech difficulty and oral incompetence may be reported.
<i>Visual acuity</i>	There are often defects in vision in one or both eyes.
<i>Cover test</i>	Strabismus is present in most cases and is usually esotropic. Where there is an associated III nerve palsy, the deviation may be exotropic.
<i>Ocular motility</i>	There is an absence of abduction of either eye and there may be other cranial nerve palsies other than VI (Spierer & Barak 2000). An occasional weakness of adduction is sometimes noted and may resemble that in internuclear ophthalmoplegia. It is postulated that this is due to an interruption of pathways in the medial longitudinal fasciculus. A disturbance of vestibular nystagmus may be demonstrated and ptosis may be noted where there is an associated III nerve palsy. Bilateral facial weakness is present with defective lid closure and a reduction of corneal sensitivity. Bell's phenomenon is retained.
<i>Associated clinical signs</i>	The associated signs may include tongue deformities, facial diplegia, deafness, dental defects, hand deformities (webbed fingers, asymmetrical fingers), feet deformities (webbed toes, clubbed feet), intellectual difficulties (variable), hypotonia and pectoral muscle hypoplasia.

Management

Correct the refractive error if present and treat amblyopia. Surgery may be required for strabismus and lateral tarsorrhaphy may be indicated for cosmesis and protection of an exposed cornea. Results of strabismus surgery are disappointing if simple medial rectus recession is undertaken (Spierer & Barak 2000). Bilateral medial rectus recessions with lateral rectus resection may be required.

Where there is a negative forced duction test, bilateral transpositions to the lateral rectus may be considered (Molarte & Rosenbaum 1990). This may be followed by graded medial rectus recessions or botulinum toxin for residual esotropia. Botulinum toxin is useful in young children before development of medial rectus contracture.

Where there is a positive forced duction test, transposition surgery and medial recti recessions are required. Maximum medial recti recessions are undertaken and stay sutures may be employed to improve alignment.

Facial surgery may involve reinnervated gracilis transplantation to re-animate the impaired sides of the face and with good results (Bianchi *et al.* 2010).

Strabismus fixus syndrome

This is a rare condition, usually congenital, where one or both eyes are anchored in a position of extreme adduction or abduction (the former is more commonly found) (Villaseca 1959).

Aetiology

Extraocular muscle fibrosis occurs where normal elastic muscle fibres are replaced by fibrous tissue (Harley *et al.* 1978).

Investigation

<i>Fundus and media</i>	Ocular ischaemia, optic nerve atrophy and central retinal artery occlusion may be seen due to secondary mechanical stretching and torsion of the optic nerve and compromised vascular supply (Mansour <i>et al.</i> 1987).
<i>Visual acuity</i>	Amblyopia will be present where there is unilateral strabismus.
<i>Cover test</i>	A marked unilateral or bilateral esotropia is commonly noted. However, the deviation may be exotropic or a vertical angle (Sharma <i>et al.</i> 2003). There is no movement on testing to fix with the affected eye.
<i>Abnormal head posture</i>	With bilateral deviations, there is a marked head posture that is often a face turn to fix with one eye.
<i>Ocular motility</i>	Marked limitations of lateral gaze are demonstrated.
<i>Forced duction test</i>	Positive.

Management

Correct the refractive error if present and treat amblyopia. Surgery is required for a cosmetic result for the existing esotropia. Surgery may involve liberal medial rectus recessions, medial rectus tenotomy, medial rectus recession on loops or use of stay sutures to anchor the eye in abduction post-operatively.

Thyroid eye disease

Thyroid dysfunction is due to disturbance of the immune system. The condition may be termed active when thyroid stimulating immunoglobulin (TSIgG) activity is most evident, and inactive, which follows active thyroid eye disease and occurs when normal thyropituitary homeostasis has returned. The inflammatory wet phase of the thyroid condition is a myogenic disorder and the following fibrotic dry phase

is a mechanical disorder. The active state (congestive, inflammatory signs of eye and orbit, conjunctival injection, chemosis, lid oedema, proptosis) is followed by the fibrotic state (restricted extraocular muscles and infiltration of mucopolysaccharides ground substance through the orbit). Pathologically, the extraocular muscle bellies are the primary sites of the disease. Inflammation of the muscle stimulates fibroblasts producing mucopolysaccharides and collagen, resulting in muscle degeneration and fibrosis. Kuriyan *et al.* (2008) state that genetic and environmental factors play a role in the pathophysiology of thyroid eye disease and may potentiate cellular and humoral-mediated inflammation within the orbit.

Lymphocytosis and mucopolysaccharide infiltration crowds the orbital spaces affecting orbital muscle tissue pressure and resilience. The confined orbit rapidly shows any increase in volume by forward movement of the globe (proptosis).

Causes of hyperthyroidism include toxic goitre, hypersecreting thyroid tumour and Grave's disease. Approximately, 25–50% of patients with Grave's disease develop eye problems. Risks include smoking, radioiodine treatment, female and variability of thyroid function (Weetman 2003, Ponto *et al.* 2010).

Investigation

<i>Case history</i>	There is often an insidious onset of diplopia. The condition is a bilateral process but motility can be unilaterally or asymmetrically impaired. Signs include upper lid retraction, lid lag, reduced frequency of blinking, weak convergence, inability to hold fixation on extreme lateral gaze, staring appearance, resistance to retrodisplacement of the eye, chemosis, oedema and injection of conjunctiva and tremor on gentle lid closure (Bothun <i>et al.</i> 2009, Dagi <i>et al.</i> 2010).
<i>Visual acuity</i>	General signs include an enlarged thyroid gland, weight loss despite a good appetite, raised body temperature resulting in sweating and heat intolerance, raised blood pressure that may lead to tachycardia, nervous agitation and tremors, irritable and emotionally labile (moody). Symptoms include diplopia associated with some degree of external ophthalmoplegia that is often worse in the morning time, pain on eye movement due to fibrosed muscles, epiphora, grittiness of eyes and general discomfort, photophobia and visual loss. Patients may have psychological consequences in relation to their disease and visual appearance (Estcourt <i>et al.</i> 2008).
<i>Abnormal head posture</i>	Visual loss in extreme cases may be due to an increase in orbital pressure with II nerve compression, from corneal exposure from marked proptosis with exposure keratitis or from induced refractive error. Presence of an abnormal head posture varies in each case. Where there is bilateral limitation of upgaze, there is chin elevation and face turns are adopted with limited lateral gaze.

Cover test

The presence of lid retraction should be noted. Central fixation may not be possible with the affected eye if a marked deviation is present. Observe recovery of large vertical heterophorias as this can indicate extended fusional reserves, if good recovery exists.

Ocular motility

Extraocular muscles are thickened markedly due to cellular infiltration. This interferes with relaxation and contraction, and slows movement. Limited movement occurs with restriction in the field of action of the affected muscle due to inadequate muscle contracture and restriction in the field opposite to the affected muscle because of inadequate relaxation (main limitation) (Fig. 15.7).

The most commonly affected muscle is the inferior rectus followed by the medial rectus, superior rectus and lateral rectus. Limited elevation is most common, followed by limited abduction, depression and adduction. The oblique muscles are also involved but to a much lesser extent. There may be retraction of the globe on downgaze.

Thyroid eye disease may be associated occasionally with myasthenia gravis and a 5% association is cited.

Lid lag (von Graefe's sign) and lid retraction may be observed. Lid lag is noted where the upper lid does not follow the eye fully when changing fixation from upgaze to downgaze. An area of white sclera will be exposed. Lid retraction is a raised upper lid due to overaction of Müller's muscle.

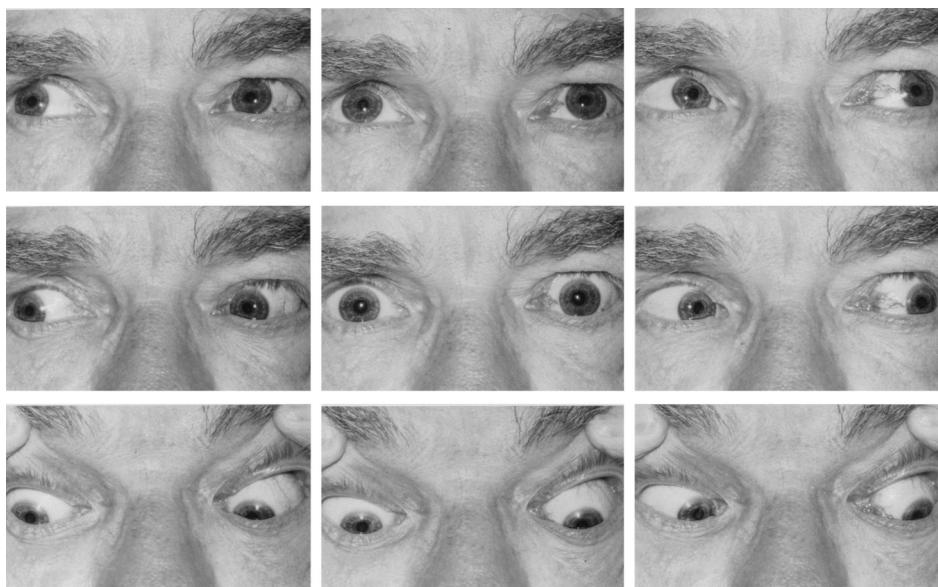


Figure 15.7 Thyroid eye disease.

There may be raised intraocular pressure on attempted elevation (Spierer & Eisenstein 1991).

The degree of ocular involvement can be graded according to severity of the condition (Werner 1969, 1977):

- 1 N No signs or symptoms
- 2 O Only symptoms
- 3 S Soft tissue involvement
- 4 P Proptosis >21 mm
- 5 E Extraocular muscle involvement
- 6 C Corneal exposure
- 7 S Sight loss due to optic nerve involvement

A Mourits' score is also available for pain, redness, swelling and impaired function (Mourits *et al.* 1997).

Convergence

This is reduced (Moebius' sign) where the medial rectus is involved.

Binocular function

Normal binocular function is often maintained with an abnormal head posture. Where there is a long-standing vertical deviation, it is not uncommon to have an extended vertical fusion range that can maintain control of large-angle vertical deviations.

Hess chart

It is important to plot the outer fields. It is necessary to ensure that the patient maintains foveal fixation of each point without moving the head. This is particularly important in cases of severe limitation of movement when documenting an accurate record of the range of ocular motility (Figs. 15.8 and 15.9).

Field of binocular single vision

This is frequently much larger than the extent of limitation seen on Hess chart (Fig. 15.10).

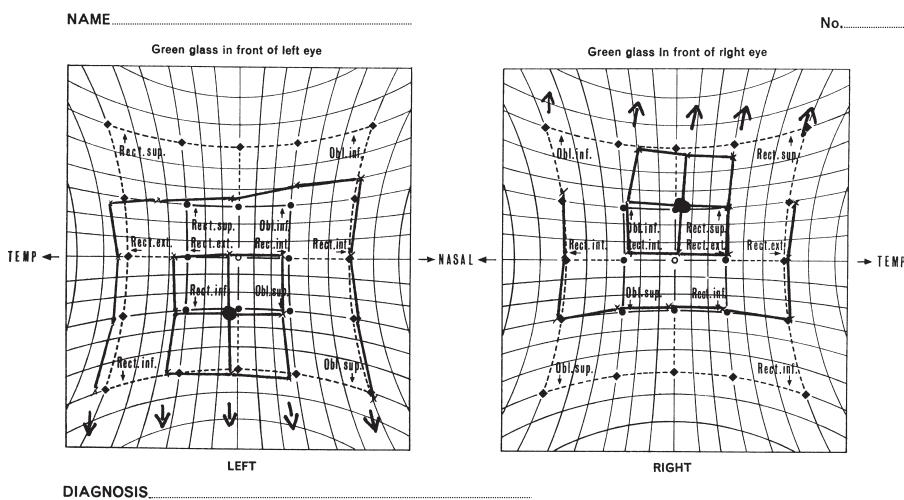


Figure 15.8 Hess chart of thyroid eye disease.

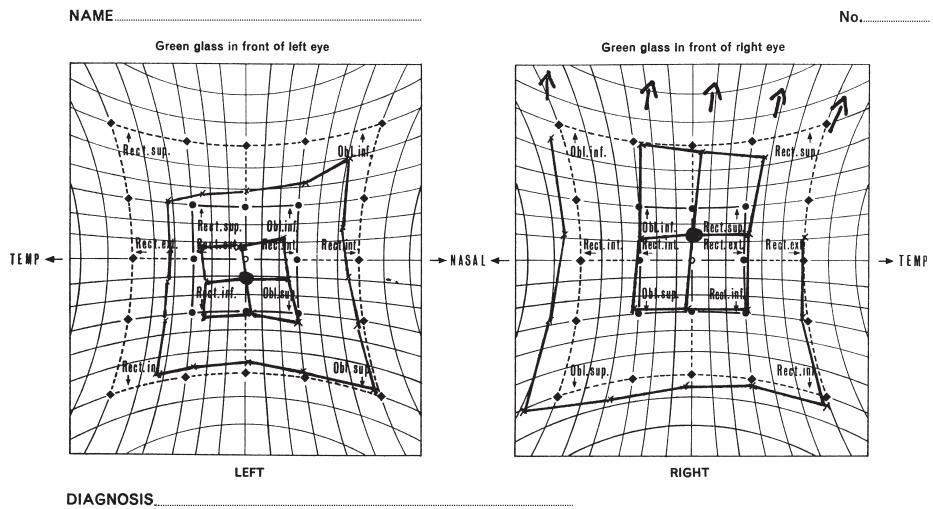


Figure 15.9 Hess chart of unilateral thyroid eye disease.

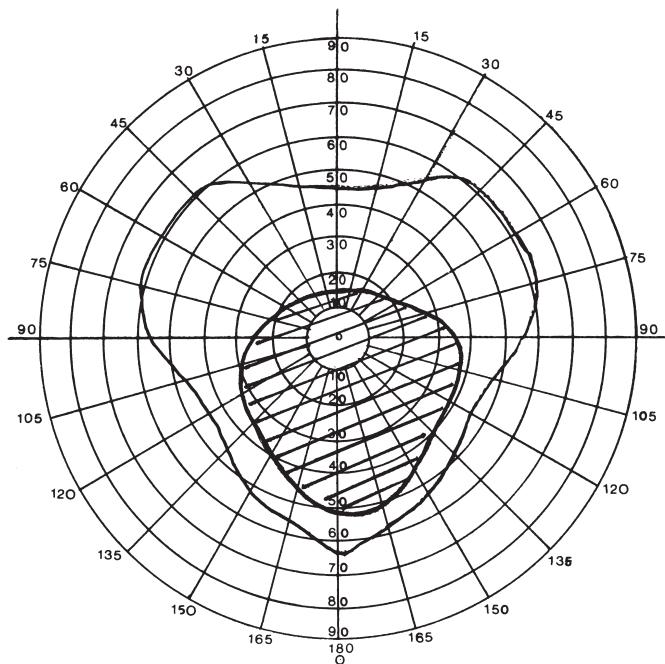


Figure 15.10 Field of binocular single vision of thyroid eye disease.

Field of uniocular fixation This is plotted if the limitation is too severe for a Hess chart to be completed (Mourits *et al.* 1994). The ocular rotations of each are measured by perimetry along six axes that correspond to the primary action of each of the extraocular muscles (Haggerty *et al.* 2005).

Ophthalmic tests

1. Measure intraocular pressure
2. Exophthalmometry
3. Fundus and media
4. Note any corneal exposure
5. Forced duction test to prove tethering.
6. Visual fields may be reduced if signs of optic nerve compression present
7. Refraction – high intraocular pressure can cause secondary hypermetropia
8. CT/MRI scan confirms thyroid eye disease by showing up thickened extrinsic muscles. A short T1 inversion recovery sequence highlights current activity within the extraocular muscles.
9. Laboratory tests include serum thyroxine T4 assay, serum tri-iodothyronine T3 assay, thyroid releasing hormone (TRH) test, radioactive iodine uptake and antibody titre test.

Management

Long-term observation of the ocular condition is required while the medical condition is treated.

Aims of treatment

1. Preserve visual acuity
2. Relief of symptoms
3. Improve cosmesis

Visual acuity

Where visual acuity is acutely compromised, an orbital decompression is indicated (Kuriyan *et al.* 2008). This may take the form of medical decompression, surgical decompression or radiotherapy. Medical decompression involves the oral or intravenous pulse administration of high dose steroids. Immunosuppressants such as cyclosporin A and azathioprine are also used. Surgical decompression involves the removal of orbital walls, often the orbital floor and the medial orbital wall. Displacement of inferior and medial recti paths may occur after decompression surgery resulting in patterns of ocular motility restriction (Abramoff *et al.* 2002). Radiotherapy to the orbital apex area reduces inflammation but its effect is not seen until approximately 6 weeks post radiotherapy and therefore is not useful where immediate decompression is required.

Prisms

Success with prisms depends on the size of deviation, the amount of incomitance and variability of the deviation and as such, frequent changes may be necessary (Lyons & Rootman 2010). If a small deviation is present, the whole vertical deviation may need to be corrected to make the patient comfortable with prisms. With a large deviation, it may be possible to manage with a considerably lower power prism because of the gradual increase in deviation and the consequent increased vertical fusion range.

Strabismus

When the ocular condition has been stable for at least 12 months and the general condition is euthyroid, *surgery* may be considered to correct the strabismic angle (Nardi 2009, Lyons & Rootman 2010). The tethered muscles are recessed on adjustable sutures (Bishop & Doran 2004, Mocan *et al.* 2007), principally the inferior and medial recti (Newman 2009). Where there are bilateral tethered muscles, the muscle that is tethered the most is put on adjustable suture. When recessing the inferior rectus, it is important to ensure that lower lid retraction does not occur (Kushner 1992).

Botulinum Toxin A may be of use with recent thyroid cases where there is some cellular infiltration, particularly with the inferior rectus and medial rectus (Wutthiphian 2008). Early injection may prevent development of fibrotic contracture and as a result may delay tethering.

General symptoms

Lid retraction may be improved by anti-thyroid drugs (propranolol and guanethidine) or surgery. Henderson's operation (a disinsertion of the smooth muscle component of the levator from the upper border of the tarsal plate) gives a good correction of established lid retraction and never produces an overaction. Botulinum toxin injection has been shown to improve upper lid retraction and reduce levator function with an adequate lid contour (Costa *et al.* 2009).

Lid swelling is a cosmetic problem. Diuretics can be used and will also reduce epiphora. Small doses of systemic steroids may also be considered.

For grittiness and general discomfort, palliative drops and artificial tears give temporary relief. If there is a conjunctival or corneal infection, antibiotic drops are needed. Tinted glasses are useful if the patient is photophobic.

Lateral tarsorrhaphy may be undertaken in severe cases. This is useful in protecting the lateral conjunctiva from irritation due to dryness caused by chronic exposure associated with proptosis (Phillips *et al.* 2010).

Orbital injuries

Orbital injuries may encompass the following:

- Blow-out fracture
- Soft tissue injury

- Supraorbital fracture
- Naso-orbital fracture
- Zygoma fracture

Blow-out fracture

A blow-out fracture occurs when a smooth contoured object of diameter larger than the orbit hits the orbit with sufficient force to force the soft contents backwards without rupturing the globe. The subsequent rise in intraocular pressure fractures one of the orbital walls, most commonly the medial and orbital floors (Song *et al.* 2009). Males present more frequently than women and causes include assault, falls, road traffic accidents, daily life accidents, sport and work-related accidents (Rowe & Crowley 2003, Gosau *et al.* 2011). The types of fracture include the following:

1. Pure:
 - Trap door
 - Linear (green stick)
 - Hanging
 - Hinged bone crack
 - Depressedor a combination of these fracture types
2. Impure

The orbital rim is also involved.

Mechanism

The limitation of ocular motility results occasionally from direct entrapment and damage to an extraocular muscle (commonly the inferior rectus). However, the limitation of ocular motility predominantly results from entrapment of orbital fascia, the septum, connective tissue and muscle pulleys, which subsequently impair extraocular muscle action (Koornneef 1979a, b, Demer *et al.* 1995).

Investigation

- Case history* Signs include periorbital ecchymosis, surgical emphysema, enophthalmos, depression of the globe, traumatic mydriasis, subconjunctival haemorrhage, hyphaema and facial asymmetry. Symptoms include diplopia (usually vertical), infraorbital anaesthesia and pain on eye movements.
- Visual acuity* Visual acuity is expected to be reduced where hyphaema or perforation of the globe has occurred.

Abnormal head posture An abnormal head posture is used to maintain binocular single vision and avoid painful areas of ocular movement. There is usually some chin elevation/depression with orbital floor fractures. There may be a face turn with medial wall fractures.

Cover test The cover test should be performed with and without any abnormal head posture and there is often a small deviation in the primary position, which is controlled to a latent deviation.

A hypotropia is noted where there is entrapment of tissue anterior to the equator with associated limitation of elevation. Hyper tropia is noted with entrapment of tissue posterior to the equator with associated limitation of depression.

Ocular motility Enophthalmos may be apparent from initial observation. There is limited elevation and depression with orbital floor fractures and limited abduction and adduction with medial wall fractures. Retraction of the globe may be noted in the position of maximum limitation (Fig. 15.11).

Diplopia may swap depending on position of gaze, due to limitations in elevation and depression. Infraorbital anaesthesia is seen with damage/bruising to the infraorbital nerve, leading to numbing of distribution of this nerve, i.e. cheek, upper lips and side of nose.



Figure 15.11 Left orbital floor fracture.

Diplopia
Binocular function
Hess chart

Field of binocular single vision
Ophthalmic tests

This reverses with limitations in opposite positions of gaze.
 There is usually a good level of binocular function maintained in the primary position with an abnormal head posture.
 This demonstrates limitations in opposite positions of gaze. It is important to plot the outer fields as slight limitations may not be demonstrable on the inner fields alone (Figs. 15.12 and 15.13).

The field is spread out across or along the centre portion (Fig. 15.14):

- Examination of fundus and media to check if the globe has been damaged while checking for retinal detachment, vitreous detachment, subluxed lens and optic nerve patency.
- Radiological examination (X-rays, CT, tomography) to pinpoint the location of the fracture.
- Measurement of intraocular pressure, particularly if hyphaema is present. Measure in primary position and in position of greatest limitation.
- Forced duction test to assess if limitation is mechanical/neurogenic.
- Measurement of enophthalmos assessed with exophthalmometer.
- Measurement of saccadic velocity.

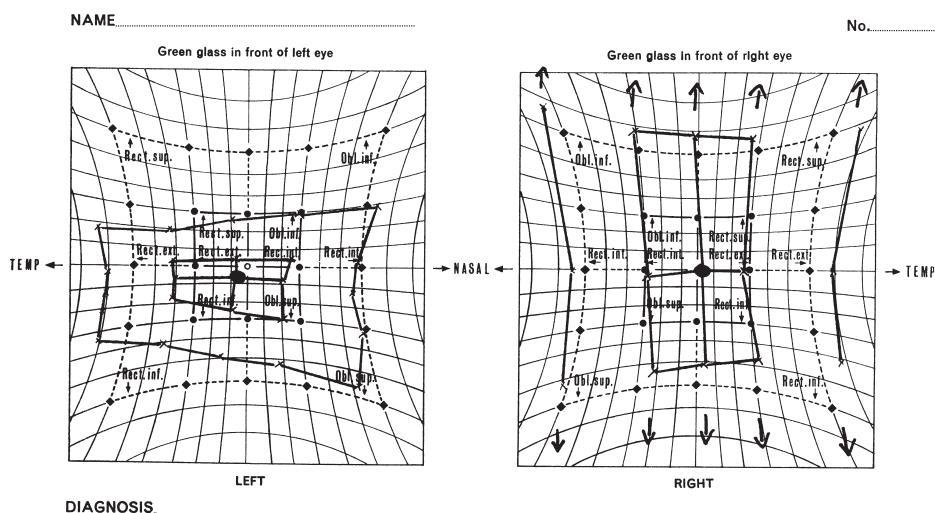


Figure 15.12 Hess chart of left orbital floor fracture.

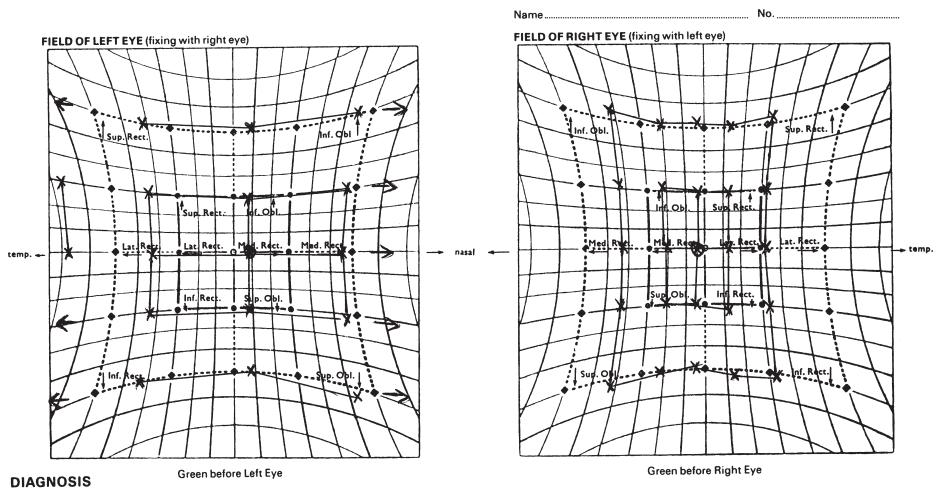


Figure 15.13 Hess chart of right medial wall fracture.

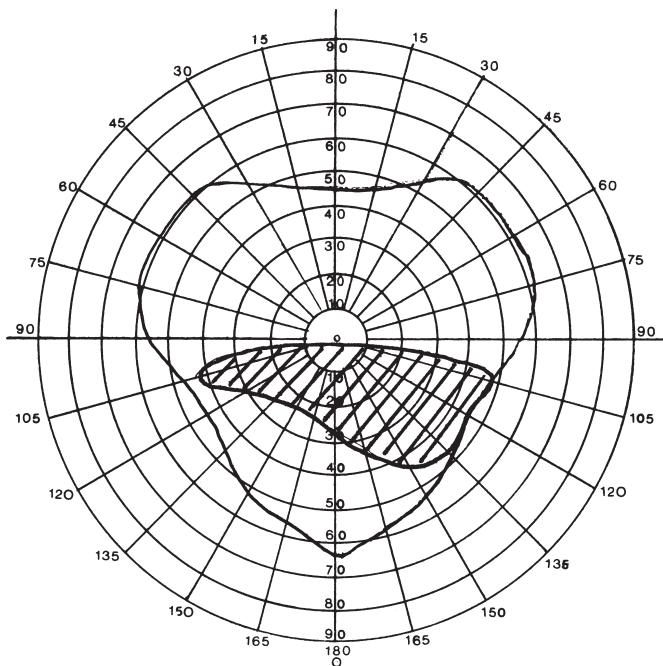


Figure 15.14 Field of binocular single vision of left orbital floor fracture.

Management

It is usual to wait for recovery in ocular motility and a wait of approximately 14 days is advisable. It was first suggested by Waddell *et al.* (1982) that patients under the age of 20 years responded less well to treatment than older patients. This was confirmed in a later study (McCarry *et al.* 1984) where it was suggested that the poorer outcome might be due to the faster formation of fibrous scar tissue in the younger age group. Further conflicting studies of childhood orbital fractures have found poor outcomes in children in contrast to the resolution of symptoms in adults (Cope *et al.* 1999) or little difference in post-operative residual diplopia whether adults or children (Leitch *et al.* 1990). There is a trend to longer recovery times with longer delays to surgery (Carroll & Ng 2010).

Many cases resolve symptomatically, despite residual limitations of ocular motility. However, patients with fractures involving a significant proportion of the orbital floor should be operated on early as enophthalmos is more common in such patients and repair of enophthalmos is more successful if performed early. Soft tissue that is damaged when trapped between bone fragments is reported as leading to fibrosis and tethering of the globe (Harris 2006). Oral antibiotics and prednisolone aid the reduction of infection and inflammation, respectively. Treatment options for orbital injuries include observation, conventional treatment (prisms, exercises and occlusion) and surgical intervention.

Early versus late surgical repair of orbital fractures has been debated (Burnstine 2002, Harris 2006, Bernardini & de Conciliis 2008). Bleeker (1967) reported that early intervention yielded better results whereas old cases were hard to deal with and responded poorly. However, Puttermann *et al.* (1974, 1991) advocated non-surgical management and suggested a period of 4–6 months wait after injury before surgery could be undertaken. At that stage conventional extraocular muscle surgery on the unaffected eye was performed to relieve residual diplopia. Dulley and Fells (1974) stated that late orbital repair had a high complication rate of both limited ocular motility and enophthalmos. They examined the optimum time for orbital repair and found it to be within 10–14 days post injury and specified criteria for surgical intervention in this time period (see below). Catone *et al.* (1988) stated that surgical intervention is recommended in patients who demonstrate residual diplopia in primary gaze and restricted ocular motility that persists after 10–14 days, the presence of enophthalmos greater than 2 mm and gross disruption of the orbital floor as confirmed by CT or tomography. Patients with severe limitation of ductions and pain, nausea and vomiting often have direct entrapment of muscle in the fracture site and early surgical repair within 1 week of injury has been shown to result in a more rapid improvement than surgery performed later (Egbert *et al.* 2000). Urgent surgery has also been advocated for patients with the ‘missing muscle syndrome’ where there is severe entrapment of the inferior rectus muscle and an absence of the inferior rectus muscle on CT study (Wachler & Holds 1998). Effective fracture repair can be undertaken up to 1 month after trauma, which allows continued observation of patients to consider improving diplopia and those at low risk of enophthalmos (Dal Canto & Linberg 2008).

Prisms may be required to join diplopia in the primary position.

Indications for surgery (Dulley & Fells 1974)

1. Diplopia not resolving
2. Enophthalmos >3 mm
3. Large fracture
4. Incarceration of tissue with globe retraction
5. Intraocular pressure increased on upgaze

Aims of surgery

1. Free the trapped tissue and repair the fracture site.
2. Correct the strabismus.

Procedures

The fracture site may be approached along the floor of the orbit or inferiorly via the maxilla sinus via a trans eyelid approach, conjunctival approach, Caldwell-Luc approach or endoscopic approach (Fernandes *et al.* 2007, Lane *et al.* 2009). Trapped tissue is freed and a silastic plate placed over the fracture site to prevent further entrapment of tissue in the fracture site.

Where surgery is indicated for strabismus, the overacting muscles of the unaffected eye may be weakened. With limited movements, recessions on adjustable sutures in the affected eye will free movement (inferior rectus if floor fracture). Where there is an associated extraocular muscle palsy, the affected muscle may be resected if a partial palsy, or if complete, weaken the overacting antagonist, consider a Faden suture on the contralateral eye to match the deficit of ocular movement, or perform horizontal recti Knapp transpositions (Hong *et al.* 2008). Where there is a generalised reduction of depression across all downward gaze positions, transposition surgery of the medial and lateral rectus muscles to the inferior rectus insertion is useful in improving downgaze, centralising the field of binocular single vision and relieving diplopia (Russell *et al.* 2006).

Soft tissue injury

This injury is due to trauma to the orbital area not causing a fracture of any bone, but resulting in damage to the orbital area and/or its contents. Orbital injuries include the following:

1. Muscle damage (Laursen & Demer 2011)
2. Lacerations (Huerva *et al.* 2008)
3. Damage to the nerve supply to the extraocular muscles
4. Retrobulbar haemorrhage causing limitation of movement and proptosis

Lid injuries include the following:

1. Lid lacerations
2. Injuries involving the lacrimal canal
3. Swelling and pseudoptosis
4. Levator damage with traumatic ptosis

Ocular signs may include any or a combination of the following:

1. Sub-conjunctival haemorrhage
2. Corneal abrasions/lacerations
3. Lens dislocation
4. Damage to iris sphincter with traumatic mydriasis
5. Hyphaema
6. Retinal detachment/haemorrhage
7. Optic nerve damage
8. Choroidal ruptures

Differential diagnosis between blow-out fracture and soft tissue injury

	Fracture with tissue entrapment	Fracture without tissue entrapment	Soft tissue injury
X-ray	Fracture	Fracture	No fracture
CT scan	Tissue entrapment	No entrapment	No entrapment
Forced duction test	Positive	Negative	Negative
Intraocular pressure	Increased on gaze opposite to fracture	No increase	No increase
Exophthalmometry	Enophthalmos	No enophthalmos	No enophthalmos
V nerve function	Infraorbital anaesthesia	Infraorbital anaesthesia	No infraorbital anaesthesia
Diplopia	Persistent	Usually resolving	Usually resolving
Ocular motility	Limitation opposite fracture site	Limitation opposite fracture site	Less marked limitation
Globe retraction	Present	Absent	Absent

Supraorbital fracture

This fracture of the orbit is caused by sharp objects going through the orbital roof.

Characteristics

1. Superior periorbital swelling and haemorrhage
2. Lid oedema
3. Supraorbital anaesthesia
4. Damage to the levator muscle and/or its nerve supply
5. Diplopia due to muscle or nerve damage, or damage to the trochlea
6. Retrobulbar haemorrhage
7. Depression of the supraorbital rim causing globe retraction
8. Cerebrospinal fluid discharge if severe

Naso-orbital fracture

This fracture is produced by direct trauma to the naso-orbital region. It usually results from road traffic accidents.

Characteristics

1. Dish-face appearance
2. Traumatic telecanthus
3. Oedema and bruising
4. Epistaxis
5. Nasal obstruction
6. Surgical emphysema
7. Damage to the tear duct and lacrimal sac

Zygoma fracture

Direct trauma to the zygoma may cause fracture and displacement of the zygoma. The bone may be displaced outwards resulting in traumatic enophthalmos, which is most noticeable after the swelling has subsided. The bone may be displaced inwards resulting in traumatic proptosis, which is most noticeable initially.

Characteristics

1. Muscle or nerve damage
2. Oedema, which may impair ocular movement
3. Infraorbital anaesthesia
4. There is a tendency to recover fairly quickly
5. Any severe head trauma could cause extraocular muscle palsies

Conjunctival shortening syndrome

There is limited ocular movement caused by contracture of the conjunctiva.

Aetiology

A long-standing deviation with contracture of the conjunctiva in marked deviations may produce this syndrome. The condition may also occur post-operatively where limitations are due to the conjunctiva not being recessed at the same time as the muscle surgery.

Investigation

<i>Case history</i>	This syndrome is associated with long-standing conditions such as thyroid eye disease, III nerve palsy, VI nerve palsy and long-standing concomitant deviations.
<i>Cover test</i>	A marked deviation is noted.
<i>Ocular motility</i>	Limited movement is demonstrable.
<i>Forced duction test</i>	Positive.

Management

Surgery is usually required in the form of conjunctival recession performed at the same time as muscle surgery. The conjunctiva may be recessed and left to re-attach. Alternatively, the conjunctiva may be resutured to the sclera in a new recessed position. It is also viable to take a flap of conjunctiva from the fornix and graft it to the recessed conjunctiva.

Retinal detachment

Retinal detachment is a separation of the main neuroretina from the pigment epithelium. The incidence of muscle restrictions, regardless of diplopia, can be high. This increases with the complexity and number of procedures (Portnoy *et al.* 1972, Sewell *et al.* 1974, Munoz & Rosenbaum 1987). In most cases, the muscle restriction is slight and often resolves with time (Kanski *et al.* 1973, Price & Pederzolli 1982, Waddell 1983, Peduzzi *et al.* 1984).

Causes of restrictions include the following (Wu *et al.* 2005):

1. Excessive or prolonged traction on a muscle(s) to position the eye for surgery.
2. The mechanical bulk of a plumb, bend or tyre under a muscle will limit its movement.
3. Formation of scar tissue around a muscle(s).
4. Disinsertion of a muscle to gain access to a certain area and then reinsertion.
5. Disinsertion of extraocular muscles.
6. Anterior migration of the sclera buckle (Wu *et al.* 2005).

Investigation

<i>Case history</i>	Symptoms are caused by post-operative oedema, reduced vision, extraocular muscle swelling and haemorrhage.
<i>Cover test</i>	Patients may have had a pre-existing strabismus. Decompensation may be due to occlusion following surgery or reduced vision.
<i>Ocular motility</i>	Following disinsertion of a muscle, it may not be reattached correctly (Kanski <i>et al.</i> 1973, Metz 1983, Waddell 1983, Wolff 1983). Encircling bands are associated with motility disturbances (Mets <i>et al.</i> 1985). Problems increase with posterior or obliquely placed buckles. Radial buckles predispose to problems (Wolff 1983, Fison & Chignell 1987).
	There may be development of adhesions and mass effect, direct muscle trauma, muscle ischaemia, muscle slippage, disinsertion, neural factors, oblique muscle inclusion or rotation, macular ectopia or gliosis.
<i>Binocular function</i>	Most patients adapt and use fusional vergences (Mets <i>et al.</i> 1985). There may be fusion breakdown and sensory deprivation, aniseikonia and anisometropia, and image distortion.

Management

Recovery is usually rapid but may take 3–6 months in some cases.

Patients may adopt an abnormal head posture. Prisms, filters and frosted lenses may alleviate diplopia both in the short and long terms.

Surgery may be required in the long term. The forced duction test should be used pre-operatively and during surgery. It may be necessary to remove the buckle, dissect adhesions and undertake a conjunctival recession. Adjustable sutures are recommended. Consider recessing overacting muscles or using a Faden suture to match the limitation of ocular movement.

Disinserted extraocular muscles can be successfully retrieved by transconjunctival surgery or orbitotomy using imaging guidance (Wu *et al.* 2005).

Cataract

It is not uncommon for diplopia to be a symptom immediately following cataract surgical procedures. This may be due to loss of suppression in pre-existing strabismus, decompensation of previous cranial nerve palsy and convergence insufficiency (Nayak *et al.* 2008). It may also relate to induced extraocular muscle imbalance due to the surgical procedure and in particular, the retrobulbar anaesthetic (Strouthidis *et al.* 2004).

Diplopia related to refractive error, lens prismatic effect and aniseikonia is frequently relieved by refractive treatment (Costa *et al.* 2006, Nayak *et al.* 2008).

Macular translocation surgery

Macular translocation surgery is a management option for occult subfoveal choroidal neovascularisation in age-related macular degeneration. It involves relocation of the macula to a new healthy portion of the retina that involves a 360° retinotomy.

Investigation

Case history Post-operatively, patients complain of diplopia, visual confusion and tilt (Mehta *et al.* 2003). Where the macula has been rotated superiorly (incyclorotation), the symptom is that of extorsion. Where the macular has been rotated inferiorly (excyclorotation), the symptom is that of intorsion.

Management

At 8–12 weeks post surgery, patients undergo removal of silicone oil with concurrent extraocular muscle surgery. The principle of extraocular muscle surgery is to counter-rotate the eyes to alleviate tilt and torsion (Eckardt *et al.* 1999, Fricke *et al.* 2000, Buckley *et al.* 2001).

Surgical procedures can include full tendon transposition of two opposite rectus muscles, crossed half tendon rectus muscle transposition, or uncrossed half tendon rectus muscle transposition. A total disinsertion of the inferior oblique muscle combined with superior oblique tuck can be used to increase incyclorotation of the globe, whereas a superior oblique tendon recession with suturing of the anterior portion nasal to the superior rectus muscle combined with inferior oblique advancement can be used in increase excyclorotation of the globe (DeClippeleir *et al.* 2006).

Following surgery, high correlations have been found between objective and subjective angles of cyclotorsion. Younger patients with small fusible amounts of torsion were found to have peripheral fusion and gross stereopsis (Sato *et al.* 2003).

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16 Myogenic Disorders

Myogenic conditions may involve individual extraocular muscles or a combination and are often bilateral conditions.

Thyroid eye disease

The wet phase of the thyroid ocular condition is a myogenic disorder that often progresses to a mechanical condition. This has been discussed in Chapter 12.

Chronic progressive external ophthalmoplegia

This is a relatively benign progressive ocular disorder that can occur as an isolated condition or as part of a more widespread clinical picture. There may be long static periods but it often progresses to a complete external ophthalmoplegia with no remissions. It occurs before the age of 30 years and is usually hereditary. In some cases, it begins in early childhood when there is a more rapid deterioration that may be complete by the age of 5 years.

Aetiology

The condition is due to primary myopathy of the extraocular muscles and often is an inherited condition (single deletions, point or multiple mutations of mtDNA: Bau & Zierz 2005).

Investigation

Case history This condition is associated with pharyngeal weakness, cerebellar ataxia, deafness, peripheral neuropathy, optic atrophy and dementia. There is often a familial tendency.

The patient may be symptom-free due to the gradual onset and symmetrical involvement of both eyes. However, the most frequent presenting symptoms are diplopia and unsightly strabismus (Wallace *et al.* 1997, Tinley *et al.* 2010).

<i>Cover test</i>	The deviation noted is usually hypotropia and exotropia.
<i>Abnormal head posture</i>	A chin elevation is adopted where there is bilateral ptosis and there is often frontalis muscle overaction. A face turn to aid fixation is adopted if a large deviation is present, particularly exotropia (Tinley <i>et al.</i> 2010).
<i>Ocular motility</i>	Limitation of movement is demonstrated, usually the elevators first and horizontally acting muscles last. Convergence is affected last of all. There is no ocular movement to caloric or vestibular stimulation.
<i>Forced duction test</i>	Ptosis may be unilateral initially, but becomes bilateral and eventually is complete with no levator function. The pupil is unaffected and there is an absent Bell's phenomenon. Positive.
<i>Associated clinical signs</i>	The condition may also affect facial muscles and muscles of mastication and occasionally the neck and shoulder muscles.

Management

Ptosis props on spectacles may be useful in cases of complete bilateral ptosis. The poor Bell's phenomenon must be taken into consideration. Anterior levator advancement or Brow suspension may be undertaken (Lane & Collins 1987). Ptosis surgery can be variable and adequate corneal cover must be ensured to prevent exposure keratitis.

Diplopia often occurs and prisms or occlusion may be used if indicated. Prisms are a useful adjunct to surgery (Wallace *et al.* 1997). Surgery is usually contraindicated as the condition is progressive and results are often unsuccessful. However, once ocular motility is stable, the aim is for a cosmetic result and to reduce an abnormal head posture. An attempt is made to move eyes from the depressed position. The procedure may involve an inferior rectus recession and superior rectus resection (Wallace *et al.* 1997) and use of stay sutures or may involve transposition of the horizontal rectus muscles to aid vertical movement (Tinley *et al.* 2010). Large lateral rectus recessions and medial rectus resections may also be considered.

Botulinum toxin is useful for small angles of manifest deviation.

Myasthenia gravis

This is a relatively rare disease where the most prominent sign is weakness of single muscles. Myasthenia gravis takes two forms. There may be ocular involvement alone or the condition may be generalised with both ocular and non-ocular features. Certain muscles are affected more frequently and many patients present initially with weakness and fatigability of the extraocular muscles, either of ptosis or diplopia or both.

Thyroid eye disease and myasthenia gravis are associated in approximately 5% of cases. Myasthenia gravis may occur at any age but is more frequently seen between the ages of 20–40 years. It may be neonatal born to myasthenic mothers but is of a remitting nature. It may be congenital with autosomal recessive inheritance. It may be juvenile that is similar to adults but affects girls more and there is good prognosis for remission (Davitt *et al.* 2000, Morris 1982). It may be of adult onset and either ocular, mild generalised, moderate generalised, acute fulminant or late severe. Characteristically, remissions occur that may last for days, weeks or years and different muscles may be affected each time.

Lambert-Eaton myasthenic syndrome is associated with antibodies directed to the voltage-gated calcium channel antibodies at the pre-synaptic nerve terminal. The condition is acquired and can be paraneoplastic and secondary to small cell lung carcinoma. Presentation is usually with lower limb weakness and autonomic symptoms (Farrugia 2011).

Aetiology

Myasthenia gravis is an autoimmune disease where there is a disorder of neuromuscular transmission due to a reduction of acetylcholine receptor sites at the motor end-plate. Antibodies are directed at the nicotinic acetylcholine receptors at the muscle surface in most cases (Farrugia 2011). This reduction occurs as a result of the patient producing antibodies to these receptor sites that in turn prevent acetylcholine from being taken up by the motor end-plate. The effectiveness of transmission is reduced and early muscle fatigue occurs as stores of acetylcholine become depleted from continued release. Eventually, there are non-specific degenerative findings of the extraocular muscles and sarcomere disorganisation (Ohtsuki *et al.* 1996). Congenital myasthenia gravis is due to mutations in proteins at the neuromuscular junction.

Investigation

<i>Case history</i>	There is often a history of fatigue involving the limbs or eyes. In particular, there is easy fatigability of legs climbing stairs, arms on combing hair, weakness of jaw, difficulty swallowing and speech and weakness of muscles of respiration. The patient may appreciate diplopia or be aware of intermittent ptosis.
<i>Visual acuity</i>	Amblyopia may develop with prolonged strabismus in childhood cases (Emoto <i>et al.</i> 2009).
<i>Cover test</i>	Decompensated heterophoria or manifest strabismus may be documented (Arblaster <i>et al.</i> 2009).
<i>Ocular motility</i>	Variable eye signs from visit to visit and at the same visit is characteristic of the condition. Variable ptosis is common (approximately 80%) and may be unilateral or bilateral (Cleary <i>et al.</i> 2008). Extraocular muscle weakness is also common and isolated single muscle paresis are typical, classically an inferior

rectus paresis. Frequently, there are multiple extraocular muscle involvements and occasionally it may mimic internuclear ophthalmoplegia. Pupil reactions are normal.

Other characteristics include Cogan's lid twitch and upper lid retraction. Investigation of ocular motility must include assessment of fatigue, saccadic velocity, ptosis and nystagmus.

Simpson test

Any fatigue on sustained lid and eye elevation is observed. There is more effect of fatigue on ptosis than extraocular muscle weakness using 60-second fatigue durations (Morris & O'Day 2004a).

Cogan's lid twitch sign

Excess innervation to the lid occurs on rapid pursuit movement from a depressed position to the primary position. Transient eyelid retraction is seen during the refixation movement (Cogan 1965).

Hess chart

Variable responses, at the same visit or from visit to visit are typical.

Tensilon test

A positive response occurs within 10–60 seconds but only lasts a few minutes. Edrophonium is given intravenously. A test dose is given followed by a brief interval and then the remainder is given in small increments. A placebo can be given first to check for functional problems. IV atropine should be available for systemic cholinergic effects.

False negatives are common especially in ocular myasthenia.

Ach receptor antibody assay

If positive, this is diagnostic but results can also be negative.

Electromyography

Reduced muscle firing.

Imaging

For thymoma or thymic hyperplasia.

Management

Medical: The condition is treated medically with anticholinesterase drugs and corticosteroids, cyclosporin A and azathioprine (immunosuppressants). Immunosuppression is not used for children (Farrugia 2011).

Thymectomy: This is often indicated as there is an autoimmune deficit (Olanow *et al.* 1982).

Prisms or occlusion: Where the patient is symptomatic, prisms or occlusion may be used to join or eliminate the diplopia.

Surgery: Where there is a residual stable deviation, surgery may be considered (Bentley *et al.* 2001, Morris & O'Day 2004b). The ocular condition must be static and stable for at least 12 months and the general condition stabilised with medical treatment (Acheson *et al.* 1991, Davidson *et al.* 1993).

Botulinum toxin: It is effective once the strabismic deviation is stable and where the angle is small (Bentley *et al.* 2001).

Ptosis props on glasses, tape, tarsal resection and brow suspension can be used/instigated depending on the severity of the ptosis (Kapetansky 1972).

Myotonic dystrophy

There is a symmetrical external ophthalmoplegia. The myotonic muscles have abnormal membranes and are therefore unable to release their contractions.

Aetiology

Primary myopathy.

Investigation

<i>Visual acuity</i>	Visual acuity may be defective where there is development of cataracts. It may also be reduced due to refractive error. Hypermetropia is common as is amblyopia (Bollinger <i>et al.</i> 2008, Aring <i>et al.</i> 2010).
<i>Cover test</i>	Strabismus is common and particularly esotropia (Bollinger <i>et al.</i> 2008).
<i>Ocular motility</i>	Progressive external ophthalmoplegia with eventual limitation of ocular motility in all directions of gaze is documented. Ptosis or pseudoptosis may be evident and there are sluggish pupils that dilate poorly with mydriatics.
<i>Associated clinical signs</i>	There may be face, neck and limb myopathy with atrophy and baldness.

Management

Ptosis props on spectacles may be of use where there is bilateral ptosis.

Surgery may be considered for the cosmetic position of eyes and ptosis but is often variable (Marshman *et al.* 1998).

Ocular myositis

Extraocular muscle inflammation occurs with resultant impairment of function depending on the extraocular muscle(s) involved (Slavin & Glaser 1982, Pollard 1996).

Investigation

<i>Case history</i>	The patient commonly complains of photophobia.
<i>Visual acuity</i>	There is reduced visual acuity. The chronic form of the condition results in raised intraocular pressure culminating in optic atrophy.

<i>Cover test</i>	The type of strabismus relates to the affected extraocular muscle and diplopia is commonly reported.
<i>Ocular motility</i>	Exophthalmos and lid oedema is evident with multiple extraocular muscle paresis.

Management

Steroids are used in the medical management of the inflammation (Pollard 1996). Surgery may be required for residual restrictive strabismus (Bessant & Lee 1995).

Kearns–Sayre ophthalmoplegia

Aetiology

There is a mitochondrial abnormality of ocular muscles.

Investigation

<i>Case history</i>	There is a childhood onset but no positive family history. Patients may complain of photophobia.
<i>Visual acuity</i>	Retinal pigment degeneration is typically responsible for reduced visual acuity. Refractive errors or optic atrophy may also contribute to reduced vision.
<i>Cover test</i>	Strabismus and nystagmus are frequently noted (Grönlund <i>et al.</i> 2010).
<i>Ocular motility</i>	Ptosis and progressive external ophthalmoplegia with eventual limitation of ocular motility in all positions of gaze is demonstrable.
<i>Associated clinical signs</i>	General signs include cardiac defects and an increased cerebrospinal fluid protein detectable on cerebrospinal fluid analysis (Petty <i>et al.</i> 1986).

Management

Prisms are of benefit where the patient appreciates diplopia.

Surgery is indicated for the alignment of the eyes and also for ptosis but there are often variable results.

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17 Craniofacial Synostoses

There are five major skull plates in the skull of a newborn including the frontal and parietal bones on either side plus the occipital bone. The frontal bones are joined by the metopic suture, the frontal and parietal bones by the coronal suture, the parietal bones by the sagittal suture and the parietal and occipital bones by the lambdoid suture. The anterior fontanelle is at the junction of the frontal and parietal bones. The posterior fontanelle is at the junction of the parietal and occipital bones and closes first after the first few months of life. The anterior fontanelle closes towards the end of the first year of life. Craniofacial anomalies occur due to the premature closure of these sutures: synostoses (Thorne 1993, Rice 2008).

Plagiocephaly

Early closure of the coronal suture occurs. This results in restricted growth of the forehead on the closed suture side. Features include a flattened forehead and compensatory bulging of the forehead on the opposite side.

Brachycephaly

There is bilateral closure of the coronal sutures. Features include a flat, broad forehead with increased height.

Scaphocephaly/dolichocephaly

Early closure of the sagittal suture results in restricted growth of the head in the side-to-side direction. Features include a narrow, elongated head.

Occipital plagiocephaly

There is closure of the lambdoidal suture resulting in a flattened back of head with bulging to the opposite side plus bulging of the forehead on the opposite

side. The ear on the closed side is often displaced backwards compared with the other side.

Craniosynostoses may be syndromic or non-syndromic. Syndromic craniosynostosis frequently involves the coronal suture and has identifiable gene mutation and chromosome aberrations. Typical syndromes include the following:

- Apert
- Pfeiffer
- Crouzon
- Saethre–Chotzen
- Craniofrontonasal dysplasia
- Unicoronal synostosis

Non-syndromic craniosynostoses frequently involve an isolated single suture and may have an underlying genetic cause.

Apert's syndrome

Features include a retruded (sunken) midface, shallow orbits, hypertelorism, beaked nose, fused fingers and toes (syndactyly) and fused cranial sutures (Jadico *et al.* 2006). Dental problems may be present. Strabismus and refractive errors are highly prevalent. Cognitive and neurological developmental abnormalities may occur. The syndrome is due to *FGFR2* gene mutation (Khong *et al.* 2007, Caça *et al.* 2009) and is an autosomal dominant inheritance.

Craniofrontonasal dysplasia

This is a rare syndrome with abnormalities of the head and face, hands and feet and other skeletal bones. There are features of coronal craniosynostosis and frontonasal dysplasia, and characteristics include ocular hypertelorism, nasal cleft, wide mouth and midface hypoplasia (Tay *et al.* 2007). It is due to X-linked inheritance.

Crouzon's syndrome

Crouzon's syndrome is the most common craniofacial syndrome and involves closure of the sagittal suture in which the features may include a hypoplastic maxilla, hooked nose, shallow wide spaced orbits, corneal exposure, amblyopia, exotropia or esotropia, refractive errors, nystagmus, coloboma, cataract and glaucoma (Gray *et al.* 2005). Dental and hearing problems may exist along with cleft lip and palate. Individuals usually have normal intelligence. It is due to *FGFR2* gene mutation and has autosomal dominant inheritance.

Pfeiffer syndrome

Typical features include bulging, wide-set eyes (hypertelorism), a high forehead, underdeveloped upper jaw and a beaked nose (Vogels & Fryns 2006). The bones

in the hands and feet may also be affected. Dental and hearing problems may occur and individuals may have short fingers and toes (brachydactyly) and webbing (syndactyly). Type 1 syndrome has normal intelligence and a normal life span. Types 2 and 3 have central nervous system defects and more severe abnormalities. Type 2 has a cloverleaf-shaped head caused by more extensive fusion of the skull sutures (Plomp *et al.* 1998). There is a trilobed appearance of the skull due to closure of the coronal and lambdoid sutures. Features include hydrocephalus, shallow orbits, proptosis and corneal exposure.

Pfeiffer's syndrome is caused by *FGFR1* and *FGFR2* gene mutations and has autosomal dominant inheritance (Vogels & Fryns 2006).

Saethre–Chotzen syndrome

Coronal suture closure occurs in this syndrome (Kress *et al.* 2006). Features include a high forehead, low frontal hairline, ptosis, wide-set eyes, broad nasal bridge and facial asymmetry. It may also have associated mild abnormalities of the hands and feet (syndactyly), toe duplication, short stature and heart defects. There may be delayed development and learning difficulties although most have normal intelligence. The syndrome is due to *TWIST1* gene mutation (Paznekas *et al.* 1998) and has autosomal dominant inheritance.

Unicoronal syndrome

There is a single suture closure of the coronal suture. It may form part of Muenke syndrome. Hearing loss and learning difficulties may occur. Strabismus is common and is frequently that of hypertropia, which relates to translocation of the superior rectus muscle pulley (Weiss & Phillips 2006).

General signs and symptoms

The following are signs and symptoms that may be noted in craniofacial synostoses syndromes:

- Full or bulging fontanelle
- Sleepiness
- Less alert
- Noticeable scalp veins
- Increased irritability
- High pitched cry
- Poor feeding
- Vomiting
- Increased head circumference
- Seizures
- Developmental delay

Ocular signs and symptoms

The following are frequently noted (Tay *et al.* 2006, Vasco *et al.* 2008):

- Reduced visual acuity
- Optic atrophy
- Proptosis
- Corneal exposure
- Globe subluxation
- Strabismus
- Hypertelorism
- Telecanthus
- Epiphora
- Refractive errors
- Amblyopia
- Optic nerve abnormalities
- Nasolacrimal obstruction

Management

Corneal protection	It is important to maintain integrity of corneal structures by maintaining an adequate tear film. There may be excessive tear production, decreased tear outflow or increased tear evaporation. Tarsorrhaphy may be indicated to protect the cornea where there are shallow orbits, eyelid malposition or corneal disease. Tear outflow may be improved by altering the eyelid position with ectropion repair and with nasolacrimal surgery. Tarsorrhaphy may be achieved by the use of botulinum toxin or with glue, tape or surgical suture.
Visual impairment	Reduced vision may be due to amblyopia with co-existent refractive errors and/or strabismus (Simon <i>et al.</i> 2009, Matta <i>et al.</i> 2010). Visual impairment may also be due to corneal exposure, optic nerve abnormalities or intracranial visual pathway abnormalities.
Strabismus	Strabismus surgery is typically delayed until after craniofacial surgery. Extraocular muscle surgery is undertaken to realign the visual axes where there is potential for fusion and for improved appearance. Prior to surgery, imaging of the extraocular muscles is recommended to determine muscle paths as globe translation with unstable muscle paths can cause pseudo alphabet patterns and incomitant strabismus.
Intracranial pressure	Shunt procedures are required to maintain control of elevated intracranial pressure and typically include ventriculoperitoneal shunts.

- Cranial reshaping Craniofacial surgery may be undertaken to reshape the skull in which space is created to allow for future growth of the brain and to decompress and protect the eye and orbit (Thanapaisal *et al.* 2010). Surgical options are dependent on which sutures are fused (Esparza *et al.* 2008) and may include the following:
- Posterior cranial vault
 - Fronto-orbital advancement
 - Monobloc advancement
 - Le Fort III midface advancement
 - Bone grafts

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18 Nystagmus

Nystagmus is a condition in which there are repetitive oscillatory movements of one or both eyes in a horizontal, vertical or rotary direction. It may be present in all positions of gaze or only in one position and is nearly always involuntary.

Nystagmus usually consists of a slow phase abnormality with a fast phase recovery and due to a disruption of gaze-holding mechanisms (vestibular system, neural integrator and pursuit systems).

Aetiology

There are a number of factors that contribute to the occurrence of nystagmus and saccadic oscillations (Thurtell & Leigh 2011). Vestibular nystagmus shows a linear constant velocity slow phase. There is an imbalance in nerve input with a resultant slow phase to the side of the lesion. An impaired neural integrator can cause gaze-evoked nystagmus with a decreasing exponential slow phase.

Saccadic movements are generated by the pulse-step mechanism and failure of the step process can result in nystagmus. The step process maintains eye position and a fault in this results in a drift of the eyes (slow phase) back towards the midline. High gain instability of slow eye movement subsystems (pursuit system) may also cause nystagmus with the nystagmus slow phase having an exponentially increasing slow phase.

Classification

Physiological

- *End point:* This is present on extreme gaze, commonly lateral gaze.
- *Optokinetic nystagmus:* A combination of pursuit and saccadic eye movements to achieve fixation of consecutive target stimulus.
- *Caloric nystagmus:* Vestibular response following stimulation or inhibition of one or more of the semicircular canals. Unilateral irrigation produces

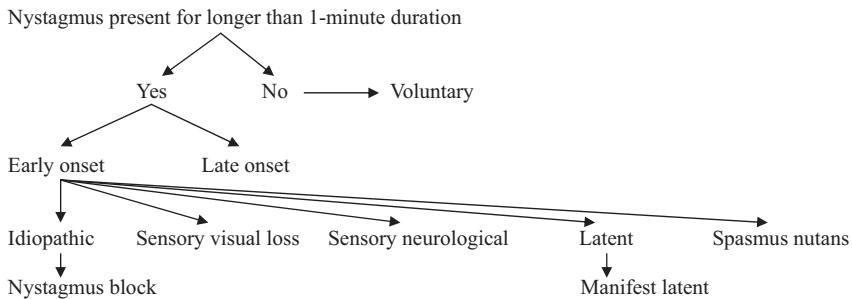


Figure 18.1 Nystagmus: early onset.

nystagmus that is horizontal, rotary or oblique, depending on the position of the head. Bilateral irrigation produces vertical nystagmus, for example COWS (cold: opposite, warm: same).

- *Rotational nystagmus:* There are fine oscillatory movements that occur during the maintenance of steady fixation.

Pathological early onset (Fig. 18.1)

- *Congenital idiopathic nystagmus:* This develops in early infancy. This is always bilateral, usually horizontal and jerky, with the fast phase to the right on right gaze and to the left on left gaze. Other waveforms include jerk nystagmus with extended foveation, pendular, pseudo-cycloid, pendular with foveating saccades and bidirectional jerk nystagmus. The waveform is the same under binocular and monocular conditions unless a latent component is present. The amplitude, velocity and frequency are similar between both eyes. The nystagmus waveform commonly changes with increasing age in three phases. The first phase is seen from 2–3 months of age and includes horizontal, wide and symmetrical swings of the eyes that are large in amplitude and of low frequency. The second phase emerges at roughly 6 months of age and consists of a pendular waveform including symmetrical and small amplitude nystagmus with a higher frequency. The third phase appears from 3 years of age and consists of jerk and slow waveforms with development of a null zone. Not all cases of idiopathic nystagmus develop to phase 3 (Cristino-Amenos & O'Connor 2007). During the development of nystagmus, adaptation occurs in which oscillopsia is suppressed and foveation periods develop to maximise visual acuity and decrease the amplitude of nystagmus.

There is often a null point between the two lateral positions of gaze and vision improves on using the null point and on convergence. An abnormal head posture is frequently adopted to make use of the null point (Hertle *et al.* 2009). The amplitude of nystagmus increases on occlusion. Nystagmus is abolished during sleep.

- *Latent and manifest latent nystagmus:* Jerky, horizontal fine nystagmus is seen when the light stimulus to one or other eye is diminished such as during

monocular viewing (Tychsen *et al.* 2010). There is an involuntary rhythmical oscillation of both eyes with the fast phase to the fixing eye. The amplitude, frequency and velocity are often asymmetrical under monocular viewing conditions.

Latent nystagmus applies to patients with binocular single vision and with no nystagmus evident with both eyes open. Manifest latent nystagmus is the term applied to patients exhibiting nystagmus of this latent type with both eyes open, but only one eye is actively used for vision due to manifest strabismus, amblyopia or ocular pathology. This is common in infantile esotropia. There may be a face turn to the side of the fixing eye.

There are four common types. The first is steady nystagmus, the second consists of horizontal square-wave jerks, the third consists of torsional nystagmus and the fourth has decelerating or linear slow-phase jerk nystagmus (Cristino-Amenos & O'Connor 2007).

- *Acquired secondary to severely defective vision:* Pendular nystagmus is seen with a jerk element on versions. Monocular visual loss may produce monocular coarse, pendular, vertical nystagmus known as the Heimann-Bielschowsky phenomenon (Smith *et al.* 1982).
- *Acquired secondary to central nervous system disease:* Infantile nystagmus syndrome may also occur in association with sensory abnormalities. Albinism is the most common (Hertle *et al.* 2009, Sarvananthan *et al.* 2009, Bagheri *et al.* 2010).
- *Spasmus nutans:* This is a rare form of nystagmus in which there is a combination of nystagmus, involuntary head nodding (titubation) and abnormal head posture (Ashurst 2002). Intermittent rapid small oscillations are seen with onset at 3–18 months, with resolution at 36 months. This is usually a benign condition with good outcome expected in one-third of cases (Gottlob *et al.* 1990, Ashurst 2002). The jerky fine nystagmus may be horizontal, vertical, rotary or a combination. The head movements do not compensate for the nystagmus and are frequently of different frequency to the nystagmus. The nystagmus can persist to the age of 12 years and co-existent strabismus develops in roughly 50%. Features of spasmus nutans can precede neurological pathology, so patients require neuro-imaging to exclude such pathology (King *et al.* 1982, 1986).
- *Nystagmus block syndrome:* The congenital nystagmus of these patients reduces or disappears on induced esotropia while fixing a distant target. During the esotropia, the nystagmus may convert to manifest latent nystagmus.

Pathological later onset (Fig. 18.2)

Particular types of acquired nystagmus have localising value and the type of nystagmus plus associated signs and symptoms can aid the determination of the site of pathology (Lee & Brazis 2006).

- *Acquired pendular (sinusoidal oscillation) nystagmus:* This is commonly the result of cerebellar or brain stem lesions due to vascular or demyelinating

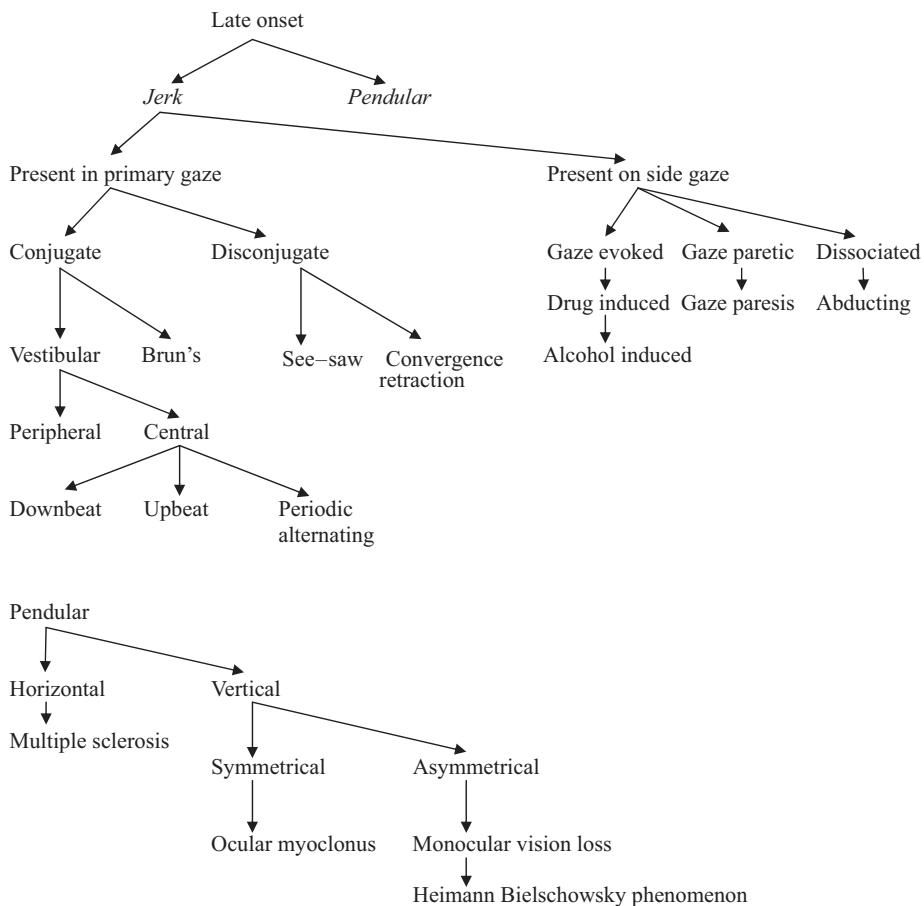


Figure 18.2 Nystagmus: late onset.

disease. The nystagmus may have horizontal and vertical components (Averbuch-Heller *et al.* 1995). A frequency of 2–7 cycles per second is seen and there may be an associated oculopalatal tremor (Tilikete *et al.* 2011).

- **Acquired jerk (slow and fast phase) nystagmus:** This may be horizontal, vertical or rotary and usually due to a supranuclear defect.
- **Vestibular nystagmus:** With peripheral disease, horizontal, jerky nystagmus is seen often with a rotary element. There may also be a vertical element in central disease due to acquired destruction of the inner ear, vestibular nerves or the cerebellar flocculus and vestibular nuclei.
- **Gaze-evoked nystagmus:** There is an inability to maintain the eyes in a lateral or vertical gaze position. The eyes drift back to the primary position, then make a correction saccade to look in the position of defective gaze. It is commonly seen during recovery of a supranuclear gaze palsy, cerebellar pathology, neuro-muscular fatigue or muscle weakness (Baier & Dieterich 2011).

- *Dissociated nystagmus*: Oscillatory movements of the eyes are noted that are dissimilar in direction, amplitude and/or speed. This may be seen where there is damage to the medial longitudinal fasciculus as in internuclear ophthalmoplegia with unilateral abducting nystagmus.
- *See-saw nystagmus*: There is alternation of the eyes where one eye elevates and intorts while the other depresses and extorts. The pattern is then repeated in the opposite direction. A frequency of 1 cycle per second is usual. This is most commonly seen in patients with bitemporal hemianopia due to large pituitary tumours or suprasellar tumours, head trauma or midbrain infarctions (Cestari *et al.* 2010).
- *Convergence retraction nystagmus*: This is classically seen in Parinaud's syndrome. On attempted upgaze, the eyes are seen to converge and retract in nystagmoid type jerk movements.
- *Downbeat nystagmus*: This is due to central nervous system lesion, often of the posterior fossa and craniocervical junction (Pierrot-Deseilligny & Milea 2005). Jerky vertical nystagmus is seen with the fast phase beating in a downward direction and with increased amplitude on downgaze (Wagner *et al.* 2009). The eyes drift up and beat down again.
- *Upbeat nystagmus*: This is due to lesions of the anterior vermis of the cerebellum or medulla (Pierrot-Deseilligny & Milea 2005). Jerky vertical nystagmus is seen with the fast phase beating in an upward direction and with increased amplitude on upgaze. The eyes drift down and beat up again.
- *Periodic alternating nystagmus*: Jerk nystagmus is seen that alters in direction every few minutes, approximately 2 minutes, with a period of oscillation of 4–5 minutes (Shallo-Hoffman *et al.* 1999). Smooth pursuit movements are commonly impaired. This may be due to pathology of the craniocervical junction, cerebellum, drugs or multiple sclerosis.
- *Brun's nystagmus*: This is due to tumours at the cerebellopontine angle such as vestibular schwannoma or arachnoid cysts (Yokota *et al.* 1993, Lloyd *et al.* 2009). There is ipsilateral large-amplitude, low-frequency nystagmus and contralateral small-amplitude, high-frequency nystagmus.
- *Drug-induced nystagmus*: This is often in the form of gaze-evoked nystagmus, most commonly upbeat jerky vertical nystagmus.
- *Alcohol-induced nystagmus*: There is often associated vertigo. It occurs with chronic disease with degenerative brain changes.
- *Voluntary nystagmus*: This purposive, self-induced nystagmus with intermittent rapid oscillations usually lasts for periods of 10 seconds. The lid flutters in association with the eye movement.

Ocular flutter/opsoclonus

This is a series of to and fro saccades. If the movement is horizontal, this is termed ocular flutter. If both horizontal and vertical, this is termed opsoclonus. A brief intermittent binocular oscillation occurs during straight-ahead fixation (Thurtell & Leigh 2011).

Square-wave jerks

Small amplitude saccades of 0.5°–5° move the eyes away from fixation and back with a 200 ms intersaccadic interval (Salman *et al.* 2008). These can occur in normal subjects and increase in darkness and with age. They are also common in cerebellar disease, Parkinson's disease and progressive supranuclear palsy.

Bobbing/dipping

There are fast downward jerks of both eyes followed by a slow drift to the midline. There are four types (Mehler 1988). The jerks may be disconjugate in the two eyes and often the eyes remain deviated for several seconds before returning to the midline. Usually occurs in comatose patients with extensive destruction of the pons.

Investigation

Case history

The age of onset, family history of nystagmus and associated symptoms should be ascertained. The medical history should be determined in regard to drug therapy, previous head injury and diseases causing central nervous system disorders (Hensil & Gurwood 2000, Moster 2001).

The patient is questioned as to whether the nystagmus is present constantly or intermittently or whether it increases on altered direction of gaze.

Associated signs/symptoms include oscillopsia, vertigo, gait and loss of acuity.

Visual acuity

Visual acuity may be normal or reduced. Visual acuity may improve with use of a null point on lateral gaze or on convergence. Latent nystagmus will result in reduced monocular acuity but with good binocular visual acuity (Spierer 1991). Patients with idiopathic nystagmus with or without albinism have visual acuity within normal ranges during infancy and idiopathic nystagmus patients show visual acuity maturation similar to normal children after 24 months whereas those with albinism do not keep pace. Nystagmus with associated optic nerve hypoplasia or retinal pathology show poor visual acuity in infancy that does not improve with age (Fu *et al.* 2011).

Abnormal head posture

Patients often have a face turn to make use of a null point to dampen the amplitude of nystagmus. The presence of an abnormal head posture appears to be independent of age, gender and strabismus. Patients are less likely to have strabismus and have better visual acuity with an abnormal head posture (Stevens & Hertle 2003).

Cover test

The pattern of nystagmus is noted. The direction of nystagmus may be horizontal, vertical, rotary or retractive and the amplitude of nystagmus may be gross, coarse or fine. The frequency of nystagmus may be fast or slow and the waveform of nystagmus may be jerky or pendular.

Jerky nystagmus involves oscillatory movements showing a fast phase in one direction and slow phase in the opposite direction.

Pendular nystagmus involves oscillatory movements that do not show a slow or fast phase. Velocity is equal in opposing directions.

The presence of a null zone should be ascertained along with the use of an abnormal head posture.

*Velocity
Ocular motility*

Figure 18.3
The pattern of nystagmus in each position of gaze is noted, recording the direction, amplitude and waveform. The direction and amplitude of nystagmus is often seen to vary on changing fixation and direction of gaze. Alexander's law states that gaze in the direction of the fast component increases the frequency and amplitude of nystagmus while gaze in the opposite direction has the reverse effect.

Convergence

The amplitude may decrease on convergence and therefore may indicate the use of a null point (Dickinson 1986).

Optokinetic nystagmus inversion of the OKN response is seen with congenital nystagmus.

*Binocular
function*

This remains for those without manifest strabismus although it may be reduced particularly if visual acuity is also impaired. It is important to assess binocular function with an abnormal head posture if present and in the position of gaze of least nystagmus (null or neutral zone).

Electrophysiology

Electrodiagnostic techniques may be used to quantify and further classify the waveform of different types of nystagmus.

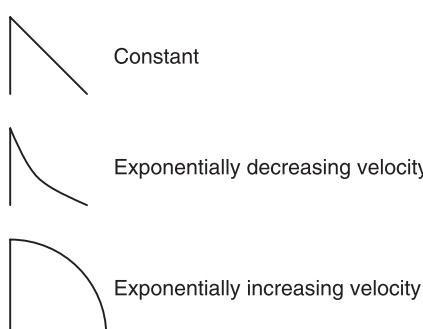


Figure 18.3 Nystagmus velocity.

Angle of deviation It can be difficult to accurately measure the endpoint neutralisation for manifest strabismus because of the overlying nystagmus movements. However, a measurement can be taken on the basis of under- and overcorrected movements. A simultaneous prism cover test is also helpful in identifying the manifest movement as distinct from the nystagmus movements.

Management

Acquired Cases

Drugs

There is a range of drugs to treat nystagmus including anti-emetics, anti-inflammatory, anti-Meniere's, anti-migrainous, anti-depressants, anti-convulsants and aminopyridines (Strupp *et al.* 2011). Specific drugs known to aid nystagmus include Baclofen, Memantine, Clonazepam, Valproate, Gabapentin and Aminopyridine (Straube 2005, Shery *et al.* 2006, Strupp & Brandt 2006).

Baclofen is usually effective in treating acquired periodic alternating nystagmus (Comer *et al.* 2006) and acetazolamide is effective in treating some cases of familial ataxia with nystagmus.

Carbamazepine has been successful in cases of superior oblique myokymia.

Prisms

These may be useful in moving eyes away from a position of nystagmus. Yoked (same strength before either eye) base-down prisms are used for upbeat nystagmus on elevation. Base-up prisms are used for downbeat nystagmus on depression.

Botulinum toxin

Botulinum toxin is useful for non-ambulant patients whereby one eye is stopped from moving by injecting retrobulbar botulinum toxin and the other eye is occluded (Dawson *et al.* 2007). Botulinum toxin may also be injected into the medial and lateral rectus muscles for cases of acquired pendular nystagmus or into all four horizontal rectus muscles (Hobson & Rowe 2009).

Surgery

Symmetrical bilateral superior rectus muscle recessions may be undertaken for chin down abnormal head posture and augmented with bilateral inferior oblique myectomy/recession or inferior rectus tenotomy. Bilateral inferior rectus muscle

recessions may be undertaken for chin elevation abnormal head posture and augmented with bilateral superior rectus recession (Hobson & Rowe 2009).

Congenital cases

Aims

- Improve acuity
- Reduce abnormal head posture

Amblyopia

Part-time total occlusion is attempted unless there is marked nystagmus. If marked, there may be an indication for use of atropine occlusion and/or penalisation. Use of a high plus lens to fog the vision will allow partial occlusion without increasing the amplitude of nystagmus (Calcutt & Crook 1972).

Non-surgical methods

Where the child sits in the classroom is important. The teacher should allow the child to use their abnormal head posture comfortably. Low-vision aids may be beneficial occasionally. Hold print close to make use of reduced amplitude on convergence.

Strabismus

Surgery should be undertaken as indicated for the strabismus. This can be combined with surgery for an abnormal head posture as explained later.

Nystagmus

Contact lenses: These may be used for correction of moderate to high refractive errors to obtain the best possible visual acuity as the lenses will move with the oscillatory eye movements (Hertle 2000). The contact lenses may also dampen some of the nystagmoid movements (Allen & Davies 1983).

Biofeedback: This system has been used to treat patients with congenital nystagmus. It may reduce but does not eliminate the oscillation (Stahl *et al.* 2000).

Prisms: These may be used diagnostically prior to surgery, or therapeutically for constant wear to determine the effectiveness of a null point (Gräf 2002). In cases with a null point on lateral gaze and abnormal head posture, prisms are placed before either eye with the apex towards the null point. In cases with reduction of nystagmus on convergence, prisms are placed before either eye base out to induce convergence.

The strength of prism is decided by checking the visual acuity level while increasing the strength of prisms.

Surgery: This is indicated if there is a marked face turn and demonstrable improvement in visual acuity with the face turn. The principle of surgery is to move the eyes conjugately and symmetrically towards the side of the face turn and away from the null point (Campos *et al.* 2003). When the patient receives the stimulus to place the eyes straight ahead, he is in fact moving into the direction of the null point and as a result, nystagmus decreases (Lee 1988, Helveston *et al.* 1991).

Commonly, the initial improvement is not maintained but reverts to approximately half way. However, patients are symptomatically improved with a broadened null zone.

Surgery is undertaken after the age of 5 years to allow for consistent abnormal head posture and other measurements.

The Kestenbaum's procedure is often the procedure of choice with beneficial effects on visual acuity and abnormal head posture (Bagheri *et al.* 2010). All four horizontal recti are moved by equal amounts: right lateral rectus resection, right medial rectus recession, left medial rectus resection and left lateral rectus recession.

A variation on Kestenbaum's procedure is Park's procedure where the four horizontal recti are moved by differing amounts (13 mm per eye; 5 and 8 mm; 6 and 7 mm).

In both procedures, the amounts can be altered to include the correction of strabismus.

Further, variations include Anderson's and Goto procedures in which two muscles are moved by recession and resections, respectively; for example null zone to left and abnormal head posture to the right:

- *Anderson's:* Right medial rectus recession and left lateral rectus recession
- *Goto:* Right lateral rectus resection and left medial rectus resection

A vertical Kestenbaum's procedure may be undertaken for vertical nystagmus with a chin up or down abnormal head posture (Schild *et al.* 2009). All vertical rectus muscles in either eye are recessed/resected (Roberts *et al.* 1996).

Surgery to dampen the nystagmoid movement intensity and improve visual acuity also includes recession (retro-equatorial) of all four horizontal rectus muscles. This procedure is reversible and does not induce severe gaze limitation or diplopia (Erbagai *et al.* 2004). Reports of beneficial improvement are mixed with some authors reporting improved visual acuity and reduced abnormal head posture in the majority of cases (Bagheri *et al.* 2005) whilst others report limited benefit (Boyle *et al.* 2006).

Artificial divergence surgery is used to dampen nystagmus based on the concept that the amplitude of nystagmus reduces on convergence. Patients must not have manifest strabismus as this procedure relies on the presence of fusional vergence to maintain alignment post-operatively. This surgery can also be combined with the Kestenbaum's procedure for a face-turn head posture (Spielmann 1982, 1994, Gräf 2002).

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19 Supranuclear and Internuclear Disorders

Disorders involving supranuclear and internuclear control pathways include (Karatas 2009):

- disorders of saccadic movements;
- disorders of smooth pursuit movements;
- disorders of the vergence system;
- gaze palsy.

The lesions resulting in supranuclear disorders are a level higher than the ocular motor nerve nuclei including lesions of the cortical centres, pathways, subcortical centres, midbrain and cerebellum. Internuclear and fascicular disorders are due to brainstem lesions.

Saccadic movement disorders

Such disorders can be produced by lesions anywhere from the frontal lobe to the paramedian pontine reticular formation (Steiner & Melamed 1984, Kernan *et al.* 1993, Shawkat *et al.* 1996).

Disorders of saccades result in abnormalities of initiation, velocity, accuracy and inappropriate saccadic intrusions. Saccadic disorders can be classified according to whether the pulse, step or the pulse-step match is inappropriate. For example, hypometric saccades relate to a deficient pulse-step signal. Where burst neurones are activated too much (excessive pulse) with a normal response from inhibitory neurones (normal step), an overshoot of the target results, which then requires a drift of the eyes (glissade) back to the target. Dysfunction of specific cell types within the brainstem reticular formation may account for various types of saccadic disorders. Slow saccades may be noted with nerve palsies or drug use. Accuracy disorders may be noted with brainstem or cerebellar disease. Disturbance of saccadic initiation with prolonged latencies and inaccuracy can be seen in degenerative and demyelinating conditions such as Huntington's chorea and multiple sclerosis (Rowe 2003).

Unilateral saccadic deficit

There is loss of voluntary movement to the contralateral side. Binocular single vision is normally maintained in the primary position but diplopia is not commonly appreciated, even looking to the affected side, as the defect is conjugate. Doll's head movement is usually intact. An object may be followed slowly if foveal fixation is maintained into the affected field. Gaze paretic nystagmus may be seen to the affected side (jerky nystagmus with fast phase to the opposite side of the lesion). There may be facial and/or upper limb paralysis on the ipsilateral side. Destructive unilateral lesions result in a unilateral contralateral saccadic deficit, which may be due to vascular or space-occupying lesions or trauma. Vascular lesions are most common such as an acute cerebrovascular accident.

Bilateral saccadic deficit

There is a complete saccadic palsy with absent horizontal and vertical saccades and no voluntary movement. Bilateral cortical lesions that may result in a complete saccadic palsy are often associated with decreased levels of consciousness.

Ocular motor apraxia

This is a congenital condition, where there is an inability to make voluntary horizontal saccades when the head is immobilised. The patient makes use of head thrusts to induce compensatory movements of the vestibular system. Typically, the eyelids are closed at the onset of the head movement in order to help break fixation. The head is thrust in the direction of and past the position of the target to be fixated. Due to the vestibulo-ocular reflex (VOR), the eyes deviate in the opposite direction. The head is moved until the eyes come in line with the target to be fixated. Once fixation is obtained, the head is moved back to a straight position making use of the normal smooth pursuit movements to maintain fixation on the target. The thrusts become less apparent with age (Rowe 1995, Harris *et al.* 1996).

The condition may also be acquired, for example secondary to inflammation and with extensive bilateral cerebral disease. Balint's syndrome is an acquired condition of saccadic palsy. There may be impaired reading and simultanagnosia, which is an inability to perceive more than one object at a time.

Huntington's chorea

This is an autosomal dominant defect with degeneration in the basal ganglia structures. There is an insidious onset of this progressive neurological disorder, usually from 30 years onwards. Marked loss of saccadic velocity is noted with abnormal control of saccades and the patients develop dysarthria and choreic movements (most noticeable in the face and distal extremities). The increase in saccadic deficits correlates with the disease severity and worsens as the disease progresses (Peltzsch *et al.* 2008). There is progressive mental deterioration in later stages.

Dementia

There is abnormal fixation with saccadic interruptions. Increased latency, impaired reflexive saccadic inhibition, hypometria and slowing of saccades are noted. Vertical saccades are affected more than horizontal saccades. With progression, there may be abnormalities of smooth pursuits with decreased gain. Saccadic deficits are due to frontal lobe dysfunction but there may also be some parietal lobe involvement. Conditions such as Alzheimer's disease and frontotemporal dementia may exhibit the above (Meyniel *et al.* 2005).

Multiple sclerosis

There is demyelination of nerve fibres with disruption of pathways in the brain stem, cerebellum and cerebral hemispheres. As a result, large saccades may show velocity changes such as increased latency and inaccuracy plus saccadic intrusions. Smooth pursuit gain can be decreased. Other ocular motility disorders may be recorded including cranial nerve palsy, internuclear ophthalmoplegia (INO), nystagmus and gaze palsies (Prasad & Galetta 2010).

Pseudo-abducens palsy

During horizontal gaze, the abducting eye may move more slowly and may reflect an increased tone of the antagonist medial rectus during lateral gaze either by abnormal convergence or impaired medial rectus inhibition (Thömke & Hopf 2001). This is an early symptom of rostral pontine and/or mesencephalic lesions such as posterior commissure lesions, pineal tumours, hydrocephalus, vascular lesions, metabolic disorders and multiple sclerosis (MS). Reading difficulties are caused by poor tracking and focus.

Smooth pursuit movement disorders

Smooth pursuit disorders are caused by lesions involving the occipito-parietal lobe and pathways to the paramedian pontine reticular formation (Leigh & Tusa 1985, Morrow & Sharpe 1995, Lekwuwa & Barnes 1996). Normal smooth-pursuit movements are replaced by series of small saccades termed 'cog-wheeling'.

Unilateral smooth pursuit deficit

Unilateral disorders are most often caused by lesions in the posterior cortical hemisphere or in the hemisphere of the cerebellum or brain stem. Cortical lesions produce loss of smooth pursuit to the ipsilateral side. This is accompanied by Cogan's sign and the patient may also have homonymous contralateral visual field defects. Cerebellar lesions produce ipsilateral smooth pursuit defects without producing the other signs found in cortical disease. Midbrain or dorsolateral pontine nuclei lesions

result in ipsilateral smooth pursuit impairment: reduced gain (Pierrot-Deseilligny & Gaymard 1992).

Cogan's sign is a tonic deviation of the eye occurring on forced lid closure and associated with lesions in the cerebral hemispheres.

Bilateral smooth pursuit deficit

Bilateral disorders are produced by diffuse posterior hemispheric, cerebellar or brainstem disease. A true smooth pursuit palsy with an absolute inability to track a moving target, in a patient with normal fixational saccades, is extremely rare.

Vergence movement disorders

Convergence paralysis, divergence paralysis and spasm of the near reflex may result from disorders of the vergence eye movement system. In certain brainstem conditions, such as INO, convergence may be absent due to involvement of the convergence fibres in the pathological lesion.

Convergence paralysis

This has been previously discussed under convergence defects in Chapter 10.

Convergence paralysis may be secondary to various organic processes such as encephalitis, diphtheria, multiple sclerosis and occlusive vascular disease involving the rostral midbrain (Guiloff *et al.* 1980, Ohtsuka *et al.* 2002). It may also occur in isolation as a sequela of a flu syndrome. Patients usually present with exotropia and diplopia for near fixation. They will have normal adduction of either eye. The deviation can be said to be concomitant across the field of gaze. Surgery to improve ocular motility and correct strabismus includes unilateral or bilateral medial rectus resection. Prior to surgery, prisms and occlusion may be used to alleviate diplopia. Botulinum toxin may also be of use for small angles.

Divergence paralysis

This may be associated with encephalitis, demyelinating disease, neurosyphilis, trauma and space-occupying lesions in and around the cerebellum (Chamlin & Davidoff 1951, Krohel *et al.* 1982, Roper-Hall & Burde 1987). The patients appreciate diplopia on distance fixation and there is an esotropia demonstrable on cover test for distance. There is normal abduction of either eye, which differentiates the condition from lateral rectus paresis (Cunningham 1972). If there are no other associated neurological factors, it is considered benign and self-limiting. Surgery to improve ocular motility and correct strabismus includes unilateral or bilateral lateral rectus resection. Prior to surgery, prisms and occlusion may be used to alleviate diplopia. Botulinum toxin may also be of use for small angles (Hoover & Giangiacomo 1993, Thacker *et al.* 2005).

Divergence insufficiency

There is intermittent or constant esotropia at distance fixation with diplopia. A reduced fusional divergence is noted (Burian 1972, Jacobson 2000). Prisms are usually beneficial and may be reduced with time. Bilateral lateral rectus resection may be considered in cases where surgery is required (Lyle 1954).

Spasm of near reflex

This involves convergence, accommodation and miosis and may be psychogenic. It is rarely due to organic disease such as trauma, dorsal midbrain syndrome, intoxication and Wernicke's encephalopathy (Dagi *et al.* 1987). This has been previously discussed in Chapter 10. The convergent deviation can become a tonic esotropia and this responds well to retro equatorial myopexy of the medial rectus muscle or medial rectus recession (Noonan *et al.* 2001, Spielmann 2006).

Gaze palsy

There is an inability to move the eyes either by saccades or smooth pursuit in a given direction. It may be caused by lesions anywhere from the cerebral cortex (where they must be profuse, involving both the frontal and occipito-parietal centres) to the paramedian pontine reticular formation.

Horizontal gaze palsy

Horizontal gaze palsies (involving both saccadic and smooth pursuit movements) result from lesions involving the sixth nerve nucleus such as vascular or space-occupying pathology or multiple sclerosis (Bronstein *et al.* 1990, Pierrot-Deseilligny & Goasguen 1984). There is often seventh nerve involvement because of its close proximity with the sixth nerve but can occur without seventh nerve involvement (Miller *et al.* 2002).

An isolated lesion of the paramedian pontine reticular formation will not result in a complete horizontal gaze palsy but a horizontal saccadic gaze palsy due to involvement of burst neurones required for saccadic generation.

Isolated horizontal gaze palsy

The sixth nerve nucleus controls all ipsilateral eye movements (saccadic, smooth pursuit and vestibulo-ocular) as it contains the abduction motoneurones as well as the internuclear neurones for adduction (Pierrot-Deseilligny 2011).

- **Congenital:** This is a familial congenital paralysis of horizontal gaze (one plus one syndrome). All horizontal conjugate eye movements, optokinetic nystagmus (OKN) and convergence are affected to a variable extent.
- **Acquired:** Lesions in the paramedian pontine reticular formation produce an ipsilateral saccadic gaze paresis and lesions in the sixth nerve nucleus produce a

complete ipsilateral gaze palsy (Bronstein *et al.* 1990). Bilateral lesions are associated with palsy of all voluntary and reflex horizontal eye movements. Acquired lesions are usually caused by multiple sclerosis (Milea *et al.* 2001).

Internuclear ophthalmoplegia

INO occurs when there is a lesion of the medial longitudinal fasciculus between the III and IV nerve nuclei (Bronstein *et al.* 1990, Pierrot-Deseilligny 2011). This causes a disconjugate deviation on conjugate gaze to the affected side with an ataxic nystagmus of the abducted eye.

Aetiology

- Multiple sclerosis
- Vascular lesions involving the basilar artery or brainstem infarction
- Space-occupying lesions of the brainstem and IV ventricle
- Infection, viral and other forms of encephalitis
- Head trauma

Bilateral INO is most commonly caused by multiple sclerosis. Unilateral INO is most commonly caused by vascular lesions due to the midline separation of the blood supply to the two halves of the brainstem. It is important to note that myasthenia gravis can mimic INO (Glaser 1966).

Investigation

<i>Case history</i>	Symptoms include diplopia and oscillopsia. Signs may include numbness and pins and needles. Previous signs include diplopia, visual loss and retrobulbar neuritis. The patient may have a history of multiple sclerosis or hypertension.
<i>Visual acuity</i>	This may be reduced if there has been a previous episode of retrobulbar neuritis.
<i>Cover test</i>	A divergent deviation is expected that may be manifest at near fixation with crossed diplopia. There may be an associated skew deviation.
<i>Ocular motility</i>	There is a limitation of adduction with abducting jerky nystagmus of the other eye. (Zee <i>et al.</i> 1987) In bilateral cases, the limitations may or may not be symmetrical. Slight limited adduction may only be apparent on repeated testing of saccades, which shows dysmetric undershooting of the adducted eye and overshooting of the abducted eye. Oscillopsia may be noted. Vertical nystagmus is frequently noted in bilateral INO.

Optokinetic nystagmus

Convergence

Asymmetric convergence

Stroud prism test

Hess chart

Field of binocular single vision

Vertical vestibular and smooth pursuit eye movements may be impaired in bilateral cases.

With unilateral INO, OKN is defective when the stripes are rotated to the side of the lesion and normal when rotated in the opposite direction.

This may or may not be present depending on the site of the lesion. Retention of convergence is noted if the lesion is posterior to the third nerve nucleus. Convergence is absent if the lesion is anterior to the third nerve nucleus and therefore interferes with that pathway. Rostral (upper) lesions are more likely to affect descending tracts that initiate convergence or the subnucleus of the medial rectus itself resulting in absent convergence. Discrete caudal (lower) lesions are more likely to spare the subnucleus of the medial rectus and descending tracts for convergence resulting in intact convergence.

The eyes are positioned so that the abducting nystagmus is barely perceptible. The target is brought toward the abducted eye along its visual axis. The position of the eye remains constant, whereas there is the stimulation to converge in the affected eye. As the target is brought closer, the amplitude of nystagmus increases.

This test is used to induce abducting nystagmus. A base-out prism (10–20 prism dioptres) is placed before the affected eye. This stimulates the eye to adduct eliciting abducting nystagmus in the other eye.

Ipsilateral limited adduction will be recorded (Fig. 19.1). Diplopia will be displaced away from the side of the affected eye (Fig. 19.2).

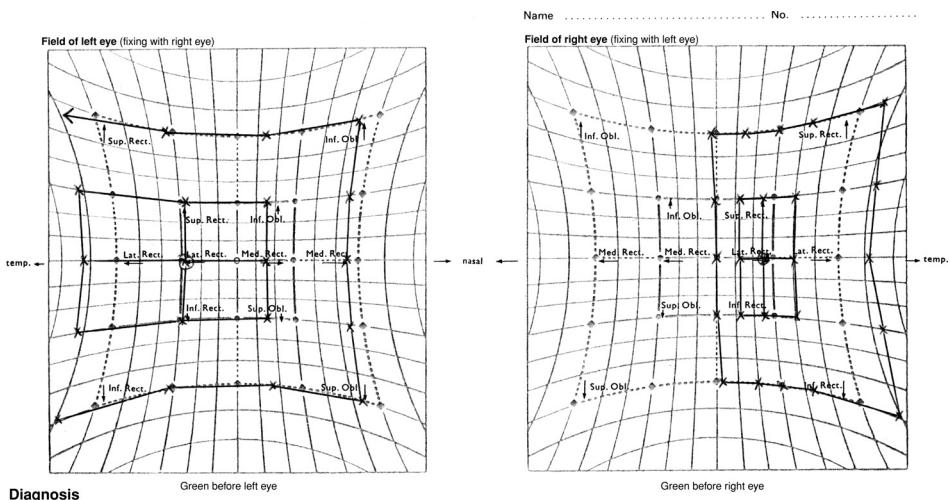


Figure 19.1 Hess chart of right internuclear ophthalmoplegia.

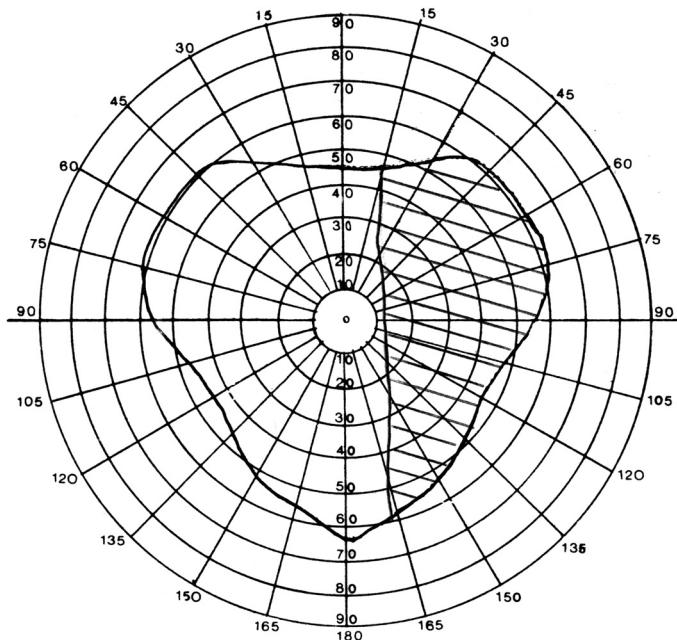


Figure 19.2 Field of binocular single vision of right internuclear ophthalmoplegia.

Management

Occlusion may be used to eliminate the symptoms of diplopia and of oscillopsia in the eye with the abducting nystagmus. Sector occlusion is often beneficial as this only occludes the medial portion of the lens to alleviate diplopia in the adducted eye position.

Prisms are often unsuccessful due to the incomitance of the deviation.

Botulinum toxin to the lateral rectus can be useful for small angle deviations. Choice of *surgery* is dependent on whether the long-standing residual ocular motility deficit is partial or complete (Roper-Hall *et al.* 2008). Lateral rectus recession and medial rectus resection may be undertaken for partial INO, whereas superior and inferior rectus muscle transposition surgery to the medial rectus is indicated for complete INO to aid improvement of adduction. A Faden procedure may be undertaken on the contralateral lateral rectus to match the deficit on lateral gaze to the affected side. Surgery frequently improves the exo-deviation and improves binocular vision. The cause is treated where possible.

WEBINO syndrome

Wall-eyed bilateral INO (WEBINO) syndrome with a marked exotropic manifest deviation and bilateral limitation of adduction. This is due to dorsal pontine or midbrain lesions affecting decussation in the medial longitudinal fasciculus such as multiple sclerosis and brain stem stroke (Chen & Lin 2007, Ushio *et al.* 2008).

INO of Lutz

INO with partial third nerve palsy. The medial longitudinal fasciculus is involved in the midbrain and abduction is impaired.

One and a half syndrome

This syndrome is produced where there is a unilateral pontine lesion that affects both the horizontal gaze centre in the paramedian pontine reticular formation and the ipsilateral medial longitudinal fasciculus, causing a combined ipsilateral horizontal gaze palsy and ipsilateral INO.

Aetiology

- Multiple sclerosis
- Basilar artery branch occlusion
- Pontine metastases of malignant tumours
- Brainstem ischaemia/infarction
- Trauma
- Pontine glioma
- Haemorrhage

Investigation

Ocular motility

There is limited adduction of either eye with limited abduction to the side of the lesion and abducting nystagmus of the other eye (Fig. 19.3).

Diplopia is often not appreciated on lateral gaze to the side of the lesion due to symmetrical horizontal gaze palsy. Diplopia and oscillopsia are appreciated to the side opposite the lesion due to limited contralateral adduction and ipsilateral abducting nystagmus.

Cover test

Exotropia in primary position – the eye opposite the side of the lesion is exotropic.

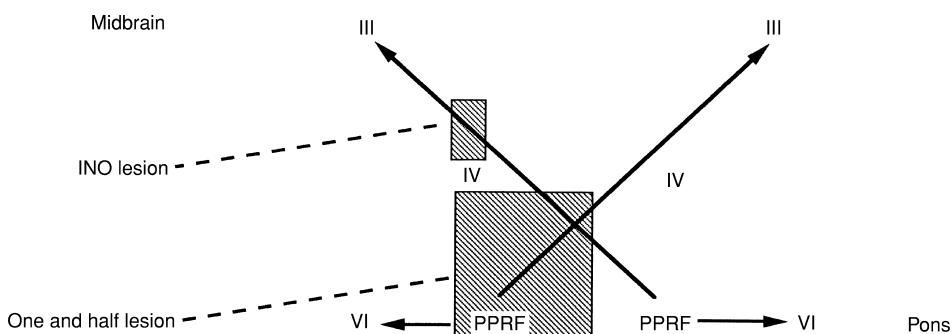


Figure 19.3 Internuclear ophthalmoplegia and one and a half syndrome – site of lesions.

Management

Occlusion may be used to eliminate the symptoms of diplopia and oscillopsia. Sector occlusion may be useful rather than full lens occlusion where symptoms do not extend across all of horizontal gaze.

Prisms are often unsuccessful due to the incomitance of the deviation.

Botulinum toxin and strabismus surgery may be undertaken for the adduction deficit and exotropia.

The cause is treated where possible.

Vertical gaze palsy

Vertical gaze palsies, whether saccadic or smooth pursuit only, or combined (more usual), commonly result from lesions in the brainstem as bilateral input is required to generate vertical gaze. The nuclei responsible for generating vertical gaze movements are connected via the medial longitudinal fasciculus and posterior commissure and vertical gaze palsies result from lesions to the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) and rostral midbrain (Bender 1980, Green *et al.* 1993, Lagreze *et al.* 1996). Saccadic innervation from the riMLF is unilateral to depressor extraocular muscles but bilateral to elevator muscles (Pierrot-Deseilligny 2011). Therefore, a unilateral lesion will result in a downward saccadic palsy. Bilateral riMLF lesions result in complete upgaze and downgaze paralysis.

Downgaze

Isolated downgaze

An isolated paralysis of downgaze has been reported in a number of cases with pretectal disease or following posterior fossa surgery. There is involvement of the riMLF bilaterally (Johkura *et al.* 1998).

Tonic downward gaze lesion

A chronic tonic downward deviation is seen associated with failure of elevation in both eyes. There may be associated convergence abnormalities and nystagmus. The condition is associated with recent bilateral thalamic infarcts or unrelieved hydrocephalus. It can also be benign in normal healthy infants (Nejat *et al.* 2008). If associated with upper lid retraction, this is termed the setting sun sign. Management typically involves lowering intracranial pressure, which results in resolution of the eye signs (Chattha & Delong 1975).

Double depressor palsy

This is extremely rare, usually congenital and sporadic (von Noorden & Hansell 1991). Causes can include inferior rectus palsy, supranuclear or secondary superior rectus contracture. Because bilateral riMLF lesions are less common than unilateral lesions, downgaze disorders are much less common than upgaze disorders.

Depression is limited in both adduction and abduction and hypertropia is seen in the affected eye. There is upper lid retraction with fixation of the unaffected eye. Differential diagnosis includes vertical retraction syndrome, congenital superior rectus fibrosis, congenital inferior rectus absence, third nerve palsy, orbital floor fracture, thyroid eye disease and surgically induced limitation.

Surgical treatment options include superior rectus recession or inverse Knapp's procedure in which the medial and lateral rectus muscles are moved downwards to the inferior rectus insertion.

Inverse parinaud's dorsal midbrain syndrome

An extremely rare condition in which there is an upper midbrain lesion affecting fibres mediating downgaze only. This is usually due to a discrete vascular lesion.

Up gaze

Parinaud's syndrome

This is an acquired palsy due to pineal gland tumour, midbrain infarction, congenital aqueductal stenosis, multiple sclerosis, arteriovenous malformation, midbrain haemorrhage, encephalitis, midbrain or third ventricle tumour, herniation of the uncus and lesions due to stereotactic surgery. There is bilateral upgaze paresis (Gregory *et al.* 2011), and when the patient attempts to look up, both eyes are seen to converge and retract into the orbit (convergence retraction nystagmus). The patient may appreciate diplopia if there is an associated III or IV nerve palsy, or skew deviation and loss of motor fusion (Gregory *et al.* 2011). The patient may also appreciate intermittent blurred vision due to papilloedema (Parinaud 1883, Lee *et al.* 1996).

The pupils may be mid-dilated with light-near dissociation (pseudo-Argyll Robertson pupil) and there may be lid retraction on downgaze (Collier's sign). Midbrain lid retraction is usually bilateral and symmetrical, but is occasionally unilateral. There is no lid lag on downgaze as in thyroid eye disease. With light-near dissociation, there is an absent pupil response to a light stimulus but normal pupil constriction occurs to an accommodative target. (When Collier's sign is associated with hydrocephalus in children, this is termed the setting sun syndrome.)

Double elevator syndrome

This is a rare acquired disorder in which there is an interruption of supranuclear input from the presumed vertical upgaze centre in the pretectum (Jampel & Fells 1968, Lessel 1975). It may be secondary to a vascular lesion. It is usually congenital but may be acquired. There is an acute onset and the patient may appreciate diplopia on upgaze due to limitation of elevation of one eye. Impaired elevation can be caused by inferior rectus restriction (Metz 1979) and supranuclear disorders (Barsoum-Homsy 1983, Thames *et al.* 1984, Ziffer *et al.* 1992).

Elevation is limited in both abduction and adduction. Abnormal head posture is usually that of a chin elevation if binocular function is present. There is usually no head posture if no binocular function. There may or may not be a manifest deviation (hypotropia) in the primary position. Rarely, fixation is with the affected eye resulting in a large secondary hypertropia. Ptosis is evident and additionally pseudo-ptosis if hypotropic in the primary position. Bell's phenomenon may be reduced and there may be associated pupil abnormalities including anisocoria, sluggish light reaction and light-near dissociation.

Differential diagnosis is with forced duction test, presence or absence of Bell's phenomenon, saccadic velocity, laboratory tests (e.g. for thyroid eye disease), neuro imaging (e.g. for fractures).

Surgical correction is generally recommended only when a vertical deviation is present in the primary position. A Knapp's procedure, transposing the horizontal recti to the borders of the superior rectus insertion, is the procedure of choice (Knapp 1969). The Knapp's procedure corrects up to 20–35 prism dioptres of hypotropia in the primary position but with little improvement in elevation. Where there is less hypotropia, there may be benefit from superior rectus resection only. An inferior rectus recession may also be considered (Metz 1981).

Upgaze and downgaze

Vertical gaze palsy

A complete vertical gaze palsy involving both upgaze and downgaze is typically due to midbrain and/or thalamic infarctions involving the riMLF, interstitial nucleus of Cajal and/or posterior commissure. A vertical gaze palsy is possible with a unilateral lesion but in which there is involvement of decussating pathways giving a functionally bilateral palsy (Alemdar *et al.* 2006).

Complete gaze palsy

Progressive supranuclear palsy

This is due to basal ganglia disease. It is a degenerative disease of the central nervous system with onset from 60 years. There is atrophy of the midbrain and dilatation of the quadrigeminal cisterns, aqueduct and third and fourth ventricles. Neuronal loss, degeneration and gliosis principally affect the brainstem reticular formation and ocular motor nuclei. It is characterised by defective downgaze first. Saccades are normally defective first, followed by smooth pursuit movements (Bhidayasiri *et al.* 2001). The condition deteriorates to complete ophthalmoplegia with absent Bell's phenomenon and convergence eventually. There are usually no ocular symptoms as the limitations of ocular movements are symmetrical. There is difficulty with swallowing and disturbance of posture and tone. The patient demonstrates mental slowing and death occurs within 5 years (Collins *et al.* 1995).

Parkinson's disease

This is a degenerative disease of the extra pyramidal motor system (Armstrong 2008), which is characterised by the triad of tremor, rigidity and akinesia. Early in the course of the disease, conjugate saccades are hypometric. Steady fixation is often disrupted by saccadic intrusions and square wave jerks. Vertical gaze is decreased and reduced blink reflex. Masked facial appearance, involuntary laughing, drooling and difficulty in swallowing indicate the progression of the disease (Repka *et al.* 1996). Diplopia is a common symptom and may be due to hypertropia, exotropia and esotropia with convergence insufficiency being a frequent finding (Lepore 2006).

Whipple's disease

This is a rare multisystem disorder, which results in weight loss, diarrhoea, arthritis, lymphadenopathy and fever (Panegyres *et al.* 2006). It causes a defect of ocular motility that mimics progressive supranuclear palsy. Vertical saccades are involved initially, progressing to loss of all eye movements. The characteristic findings include pendular vergence oscillations and concurrent contractions of the masticatory muscles (oculomasticatory myorhythms). Diagnosis is confirmed by polymerase chain reaction analysis of an intestinal biopsy sample (Averbuch-Heller *et al.* 1999). Treatment is with antibiotics.

Oculogyric crisis

A tonic vertical or horizontal deviation occurs. This can be overcome with the smooth pursuit vestibular reflex but the eyes then return to the tonically deviated position. It is seen with post-encephalitic Parkinsonism and neuroleptic toxicity. There may be obsessional fixation of individual thoughts. Symptoms resolve with reduction or cessation of drug therapy after which toxicity levels drop (Leigh *et al.* 1987).

Roth Bielschowsky phenomenon

Loss of saccadic and pursuit movements occur and at times include vertical movements. Eventually, total paralysis of eye movement develops. It is due to disruption of occipital and frontal systems with bilateral hemispherical and brainstem defects involving both the saccadic and smooth pursuit pathways. The vestibular system may remain intact.

Optokinetic movement disorders

OKN is impaired by any lesion involving the saccadic or smooth pursuit pathways and may also be impaired by any disruption of the neural integrators such as with cerebellar disease.

Vestibular movement disorders

The VOR and OKN may be affected by vestibular system lesions. Peripheral vestibular disorders due to imbalance of semicircular canal inputs may cause nystagmus, dynamic imbalance with lower gain for horizontal head rotation, loss of velocity determination with reduced VOR and OKN and skew deviation. There may be head tilts due to perceived tilts of the environment. Cerebellar lesions may also affect the VOR gain but impair smooth tracking and gaze holding (neural integrator function). Symptoms of vertigo and oscillopsia may be noted. The VOR may be involved in pontine lesions but often remains intact in rostral brainstem lesions as ascending fibres from the cerebellum are below the level of the lesion (Rowe 2003).

Brainstem syndromes

Discrete lesion of paramedian pontine reticular formation

The paramedian pontine reticular formation contains the nucleus pontis centralis caudalis and is responsible for saccadic generation via burst neurones. There is loss of saccadic and quick phase nystagmus to the side of the lesion. Vertical saccades are misdirected obliquely away from the side of lesion.

Nuclear sixth nerve

An ipsilateral horizontal gaze palsy occurs as the VI nucleus controls horizontal gaze. This is rare as usually larger lesions occur, affecting medial longitudinal fasciculus and paramedian pontine reticular formation also.

Fascicular sixth nerve

Various syndromes occur depending on the site of lesion within the brain stem and the structures involved (Silverman *et al.* 1995). This results in ipsilateral abduction paralysis, facial palsy, Horner's syndrome, facial analgesia and peripheral deafness. Commonly, this is due to vascular episodes.

Millard-gubler syndrome

This syndrome consists of VI nerve palsy, ipsilateral VII nerve palsy, contralateral hemiparesis.

Raymond's syndrome

This syndrome consists of VI nerve palsy and contralateral hemiparesis.

Foville's syndrome

This syndrome consists of horizontal gaze palsy, V, VII and VIII nerve palsies and ipsilateral Horner's syndrome.

Nuclear third nerve

Pupil, extraocular muscle and levator function are described as placed in a rostral-caudal order in the midbrain (Saeki *et al.* 2000). The inferior rectus and inferior oblique nuclei are more rostral than the medial rectus, superior rectus and levator nuclei. The following subtypes may occur:

- Unilateral III with contralateral superior rectus palsy and bilateral ptosis
- Bilateral III with spared levator function
- Bilateral total III
- Bilateral ptosis
- Bilateral medial rectus palsy
- Isolated medial rectus, inferior oblique and inferior rectus palsy

Commonly due to infarction, demyelination and/or metastatic tumour.

Fascicular third nerve

Syndromes consist of:

- III nerve palsy and contralateral hemiplegia with involvement of corticospinal tracts;
- III nerve palsy with contralateral ataxia and intention tremor with involvement of red nucleus.

These are commonly due to vascular lesions (Schmidt 2000).

Nothnagel's syndrome

There is a lesion in the superior cerebellar peduncle producing an ipsilateral III nerve palsy and cerebellar ataxia.

Benedikt's syndrome

There is a lesion in the region of the red nucleus resulting to an ipsilateral III nerve palsy and contralateral hemitremor.

Weber's syndrome

There is a lesion in the area of cerebral peduncle, resulting in ipsilateral III nerve palsy and contralateral hemiparesis.

Claude's syndrome

This syndrome consists of features of Benedikt's and Nothnagel's syndrome.

Nuclear/fascicular fourth nerve

This may be due to nuclear aplasia, arteriovenous malformation, demyelination, haemorrhage, ischaemia/infarction or trauma.

Differential diagnosis of nuclear and fascicular is almost impossible. It may be associated with central Horner's syndrome and relative afferent papillary defect.

Medulla lesions

Disease affecting vestibular nuclei and nucleus prepositus hypoglossi results in vertical imbalance such as nystagmus (upbeat, torsional and sometimes horizontal with gaze-evoked component), vertigo or skew deviation and impairment of gaze holding for all conjugate eye movements.

Wallenberg syndrome occurs with lateral medullary infarction (Dieterich & Brandt 1992). Features include skew deviation with ipsilateral hypotropia, cyclo-deviation (lower eye more exocyclorotated) and ipsilateral head tilt (ocular tilt reaction), ipsilateral facial pain, Horner's syndrome, limb ataxia, dysarthria, dysphagia, contralateral trunk and limb pain).

Skew deviation

This is an acquired vertical misalignment of the visual axes caused by a disturbance of supranuclear function (Keane 1975, Brandt & Dieterich 1993). Disease of the vestibular system (otolith organs, nuclei or nerve) can cause a skew deviation, as can cerebellar and brainstem disease (Schlenker *et al.* 2009). A disturbance of tonic vestibulo-ocular coordination is considered to be a primary factor in the aetiology of skew deviations. The deviation is more common in unilateral lesions than bilateral lesions and the eye ipsilateral to the lesion is usually the hypotropic eye. Rostral (upper) lesions more commonly cause ipsilateral hypertropia whereas caudal (lower) lesions cause contralateral hypertropia. Diplopia is often present but no particular extraocular muscle is involved. There may be a torsional element. The deviation may be constant in all positions of gaze or variable, when the vertical height reverses on laevo- and dextroversion. Alternating skew deviation, in which the hypertropic eye occurs on the side of horizontal gaze direction, is associated with lesions of the cervicomedullary junction or cerebellar pathways. When incomitant, the deviation may be differentially diagnosed from a vertical extraocular muscle palsy by the coexistence of signs of central neurological dysfunction. In addition, the symptom of torsion can be reviewed: exocyclotorsion is expected in fourth nerve palsy, whereas incyclotorsion is expected in skew deviation. Furthermore, persistence of the vertical deviation is seen in fourth nerve palsy

when the patient is supine, whereas reduction in ocular torsion and vertical deviation from standing to supine positions is indicative of skew deviation (Parulekar *et al.* 2008).

Management is aimed at the vertical deviation to alleviate symptoms of diplopia. This may be achieved with prisms or occlusion in the acute stage. Where the deviation persists long term, surgery may be undertaken. An alternating hypertropia may respond to bilateral inferior rectus resections. A non-alternating hypertropia may respond to inferior rectus resection and/or superior rectus recession (Siatkowski *et al.* 2003).

Ocular tilt reaction

In a normal tilt reaction, static lateral head tilt causes sustained low-amplitude counter-roll of the eyes and a small vertical misalignment, so that the lower eye is a little higher than the upper eye. This has the effect of realigning the axis between the eyes with the horizontal plane. In pathological damage, there is a triad of skew deviation, ocular torsion and head tilt. Disruption to the interstitial nucleus of Cajal and its inputs and projects can cause an ocular tilt reaction (Le 2007).

Ocular investigation

<i>Case history</i>	General observations should include gait, paralysis or weakening of function, the level of patient's consciousness and mental alertness, and speech. Many patients may be disorientated and may feel generally unwell. Symptoms can include vertigo, pain, loss of sensation or tingling, reading difficulties, diplopia and oscillopsia. Observations may be made while taking the case history such as the presence of a head posture, pupil size, lid position and ocular posture.
<i>Visual acuity</i>	This may be reduced if papilloedema or any lesion of the optic nerve affecting the macular fibres in particular is present. Optic neuritis may be present in cases of multiple sclerosis. Contrast sensitivity and colour vision may also be impaired, particularly with visual pathway involvement.
<i>Cover test</i>	Usually binocular single vision is present with a latent deviation in the primary position. However, exotropia or esotropia may be seen with convergence or divergence paralysis. There may be a skew deviation or vertical misalignment in some unilateral conditions such as INO, one and a half syndrome and double elevator syndrome. Nystagmus may be present in primary gaze or gaze away from primary position.
<i>Ocular movements</i>	All eye movement systems should be assessed. Saccadic intrusions, square wave jerks and fixation stability are frequently noted. Ocular dysmetria may be noted with defective saccades. Dysmetria indicates overshooting of saccades, hypometria

indicates undershooting. Ocular flutter is a disturbance of position maintenance system with unsteady movement on fixation.

Smooth pursuit movements should be assessed for conjugacy and smoothness of movements as these can be replaced by a series of small amplitude saccades (cog-wheel movements).

When assessing the integrity of vergence movements, the convergence and divergence mechanisms should be included. Unilateral or bilateral gaze deficits should be differentiated.

Nystagmus is common in many lesions, particularly if the vestibular apparatus is involved. Note the speed, amplitude and position of the greatest and the least amplitude, whether movements are jerky and if so, the direction of the fast phase. Nystagmus may only be present on versions.

Testing OKN assesses the integrity of both saccadic and smooth pursuit pathways. It is tested in both directions plus horizontally and vertically. Responses are compared on each occasion. Any difference in response tested in either direction is indicative of abnormality.

Doll's head/head rotation may be used to assess the integrity of the vestibular pathway. Caloric stimulation may be used for some, particularly semi- or unconscious, patients. Instillation of cold water into one ear results in eye movements in the opposite direction. Instillation of warm water into one ear results in eye movement towards the same direction (COWS; cold: opposite, warm: same).

These patients usually demonstrate normal binocular single vision except in cases of prior existing strabismus. In conjugate deviations, diplopia is rarely appreciated as these deviations are symmetrical. Diplopia will be appreciated in asymmetrical deviations.

In line with routine orthoptic evaluation, the angle of deviation must be measured with the appropriate test. Defective ocular movements may be recorded on a Hess chart and areas of diplopia recorded on a field of binocular single vision.

Binocular function

Angle of deviation

Ophthalmic investigation

Visual fields Post-chiasm lesions result in homonymous hemianopia or quadrantanopia. Pituitary lesions may lead to bitemporal hemianopia. Fusion is difficult due to this as there is no overlap of the visual fields. Field defects may cause difficulty in testing refixation movements. If the target to be fixed falls within a scotoma,

there may be an apparent defect of the saccadic system as no refixation movement will take place. However, on OKN testing the saccadic system may be intact. The type of field defect is a good localising sign for many lesions affecting the visual pathway at any point along its course.

Forced lid closure

This can help in the diagnosis of vertical gaze anomalies. In dorsal midbrain syndrome, there is no upgaze movement but elevation is present on lid closure indicating the integrity of the superior recti muscles and infranuclear pathway. In some parieto-occipital lesions, the eyes may deviate to one side. This is known as Cogan's sign.

Sensory function

Involvement of the V or VII cranial nerves may result in impaired corneal sensation, face sensation and function.

Management options

As evident from the discussion of these disorders, the prognosis for many conditions is poor, particularly those with degeneration. The primary aim following orthoptic investigation is to render the patient asymptomatic. Many conditions will have conjugate deviations and as a result will not appreciate diplopia. For those with asymmetrical deviations, diplopia is a frequently appreciated symptom. An abnormal head posture may be sufficient to adapt to a small area of diplopia on gaze away from primary position. Reading strategies may also be of use such as using a typoscope to narrow the field of view when reading text. Prisms may be tried but the incommittance of the deviation often makes the use of prisms difficult. Occlusion may then be considered. Occlusion (total or sector) is also useful where there is dissociated nystagmus and oscillation. Convex lenses or miotics may be prescribed for those with associated pupil and accommodative anomalies. It may be necessary to alter medication dose if this is contributing to the presence of the ocular motility deficit.

Botulinum toxin may be of use for small angle deviations and residual deviations. Surgery as for other forms of strabismus may only be considered in a stable deviation. Concomitant deviations in association with congenital supranuclear disorders may be corrected in line with normal indications, for example infantile esotropia with ocular motor apraxia. For incomitant deviations, there are a variety of different options relevant to the type of ocular motility deficit. Kestenbaum or Anderson procedures are indicated for horizontal gaze palsy with significant face turn as per guidelines for surgery in cases of nystagmus with face turn (Coats *et al.* 1998, Campos *et al.* 2003). Vertical rectus muscle transposition surgery may be required for limited adduction (Snir *et al.* 2005b). Vertical Kestenbaum procedure or bilateral Knapp's procedure is indicated for cases of upgaze palsy. Where augmentation is indicated for such surgery (such as in the presence of a positive forced duction test), additional inferior rectus recession and/or superior rectus resection may be indicated (Buckley & Holgado 2004). Vertical Kestenbaum procedure or bilateral

inverse Knapp's procedure is indicated for cases of downgaze palsy. For double elevator palsy, recommended surgery is a unilateral Knapp's procedure with additional inferior rectus recession, if there is a positive forced duction test (Kocak-Altintas 2000, Snir *et al.* 2005a). For double depressor palsy, recommended surgery is a unilateral inverse Knapp's procedure with additional inferior oblique disinsertion, if there is a positive forced duction test (Maurino *et al.* 2001). Prior to such surgery, yoked prisms may be used diagnostically to determine the effect on the abnormal head posture.

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SECTION IV

APPENDICES

Diagnostic Aids

Investigation considerations

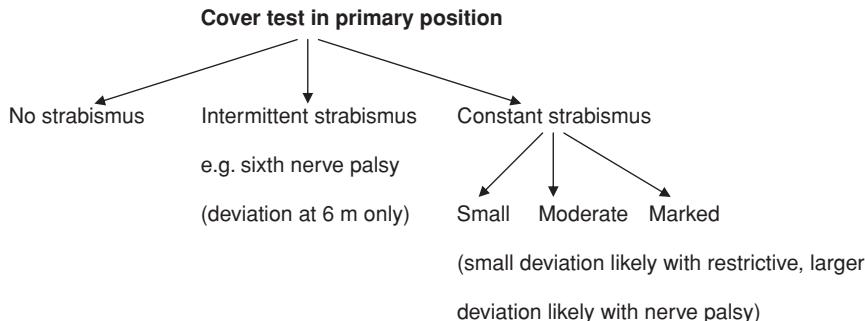
Observations	Lids, pupils, abnormal head posture, eye position, facial asymmetry, gait, photographs, general health
Patient information	Previous ocular history, mode of onset, signs and symptoms, family history, general health, birth history, referral pathway, age at onset, frequency, impact of condition, serial records
Visual function	Visual acuity, visual behaviour, contrast sensitivity, colour vision, visual field
Refraction	Cycloplegic, objective, subjective
Ocular movements	Saccades, smooth pursuit, vestibulo-ocular reflex, vergence (convergence, divergence), ductions, versions, Bielschowsky head tilt test
Diplopia	Field of binocular single vision, Hess/Lees chart, diplopia chart, postoperative diplopia test, monocular diplopia, Worth's light
Accommodation	Near point, facility, amplitude
Binocular function	Simultaneous perception, superimposition, sensory fusion, motor fusion, stereopsis, control (binocular visual acuity, controlled binocular acuity), suppression (area, density, central)
Angle measurement	AC/A ratio, prism adaptation, diagnostic occlusion, postoperative diplopia test, +3 DS lenses, position of gaze, torsion, field of fixation
Ophthalmic	Fundus and media, intraocular pressure, exophthalmos, bloods (glucose, thyroid stimulating hormone, acetylcholine receptors, erythrocyte sedimentation rate), blood pressure, ocular dimensions, visuscopy, Tensilon test, ice-pack test, slit-lamp test, forced duction test

Electrodiagnostics Electroretinogram (ERG), electro-oculography (EOG), visual evoked potential (VEP)

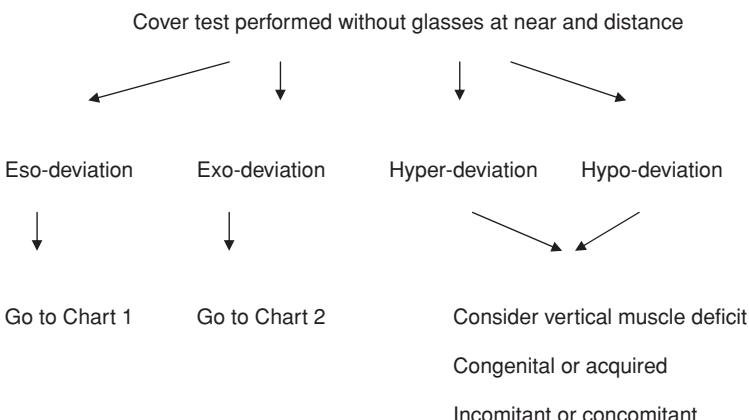
Imaging X-ray, computed tomography (CT), magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), Positive emission tomography (PET)

Cover test pointers

- Compare near and distance fixation
- Compare with and without abnormal head posture; why has the patient developed a head posture?
- Latent deviation, intermittent manifest or constantly manifest
- Eso-, exo-, hyper- or hypo-deviation (watch lids to help detect vertical deviation)
- Primary and secondary deviation
- *Diplopia*: Type, is it relevant to the deviation seen?
- Nystagmus, pupil anomalies



Cover test results



Ocular movement pointers

Smooth pursuit assessment from primary position into 8 cardinal positions:

- Are the movements symmetrical and smooth in either eye?
- Underactions, overactions, restrictions – compare ductions and versions
- A or V patterns
- Nystagmus, lid or pupil changes
- Enophthalmos or exophthalmos
- *Diplopia*: Type, is it relevant to the deviation seen in each position of gaze?

Saccadic assessment

- Compare horizontal and vertical saccades
- Compare either eye for symmetrical movement
- Hypo or hypermetric movements, under- or overshooting

Vergence assessment

- *Convergence to nose*: Do both eyes move symmetrically?
- *Divergence*: Do both eyes relax out from converged position to distance fixation?

Ocular movement results

Limited abduction

Unilateral		Bilateral	
+ve FDT	-ve FDT	+ve FDT	-ve FDT
Duane's retraction syndrome (types A and B)	Acute sixth nerve palsy	Duane's retraction syndrome (types A and B)	Infantile esotropia
Chronic sixth nerve palsy	Slipped lateral rectus muscle	Chronic sixth nerve palsy	Nystagmus block syndrome
Medial wall blow-out fracture		Thyroid eye disease	
Iatrogenic (tight medial rectus)		Chronic progressive external ophthalmoplegia	

Limited adduction

Unilateral		Bilateral	
+ve FDT	-ve FDT	+ve FDT	-ve FDT
Duane's refraction syndrome (type C)	Internuclear ophthalmoplegia Slipped medial rectus muscle	Duane's refraction syndrome (type C)	Internuclear ophthalmoplegia Third nerve palsy Myasthenia gravis
Thyroid eye disease	Myasthenia gravis	Thyroid eye disease	
Iatrogenic (tight lateral rectus)		Chronic progressive external ophthalmoplegia	

Limited elevation

Unilateral		Bilateral	
+ve FDT	-ve FDT	+ve FDT	-ve FDT
Chronic third nerve palsy	Double elevator palsy	Thyroid eye disease	Dorsal midbrain syndrome (Parinaud's)
Thyroid eye disease	Acute third nerve palsy	Chronic progressive external ophthalmoplegia	Tonic downward gaze palsy
Orbital floor fracture	Concomitant hypotropia		Bilateral high myopia
Vertical Duane's retraction syndrome	Heavy eye syndrome		
Brown's syndrome			
Double elevator palsy			
Orbital myositis			
Retinal detachment surgery			

Limited depression

Unilateral		Bilateral	
+ve FDT	-ve FDT	+ve FDT	-ve FDT
Chronic fourth nerve palsy	Double depressor palsy	Thyroid eye disease	Inverse dorsal midbrain syndrome (Parinaud's)
Thyroid eye disease	Acute fourth nerve palsy	Chronic progressive external ophthalmoplegia	
Orbital floor fracture	Concomitant hypertropia		
Double depressor palsy			
Orbital myositis			
Retinal detachment surgery			

Maintenance of binocular vision/alleviation of diplopia

Conservative

- Abnormal head posture
- Prisms
- Occlusion
- Bangerter filters

Intervention

- Botulinum toxin
- Surgery

Angle of deviation pointers

- Measure angle of deviation at near and distance fixation, with and without abnormal head posture.
- Is there a primary and secondary angle of deviation? If yes, could indicate restrictive condition or recent onset cranial nerve palsy.
- Measure angle of deviation in cardinal positions of gaze.
- Where is the maximum angle of deviation?
- Does this coincide with the patient's perception of maximum symptoms?

Diagnostic Considerations

Always consider whether the ocular motor problem could fit a category of motility impairment. Remember to look for associated signs such as lid and pupil changes:

- *Neurogenic*: III, IV or VI cranial nerve palsy or combination
- *Mechanical*: Brown's syndrome, Duane's retraction syndrome, thyroid eye disease, orbital fractures
- *Myogenic*: Myasthenia gravis, chronic progressive external ophthalmoplegia, extraocular muscle inflammation
- *Nystagmus*: Check direction, waveform, amplitude, frequency and velocity
- *Internuclear*: Internuclear ophthalmoplegia, one and a half syndrome
- *Supranuclear*: Saccadic deficit, smooth pursuit deficit, vergence paralysis, gaze palsy, skew deviation

Remember to work through the most likely diagnoses first.

Management considerations

Observations	Refractive adaptation, post surgery adaptations, abnormal head posture
Orthoptic exercises	Recognition of diplopia, improve relative vergences, control of deviation, improve fusional amplitude, eliminate suppression
Optical	Prisms for adaptation, join diplopia, exercise fusional vergences, alleviate abnormal head posture. Lenses to induce or minimise accommodative convergence, bifocals, correction of refractive error, decentration for prismatic effect
Amblyopia	Total form occlusion, total light occlusion, partial occlusion, atropine, penalisation
Occlusion	Elimination of diplopia
Advice	Low vision aids, abnormal head posture
Botulinum toxin	Diagnostic procedure, long-term therapeutic procedure, postsurgery enhancement, postoperative diplopia assessment
Medical drugs	Miotics, corticosteroids, tensilon, mydriatics
Surgery	Strength muscle function, weaken muscle function, transpose muscles to facilitate gaze, selectively weaken function to match deficit
Referrals	Low vision services, medical services, education services

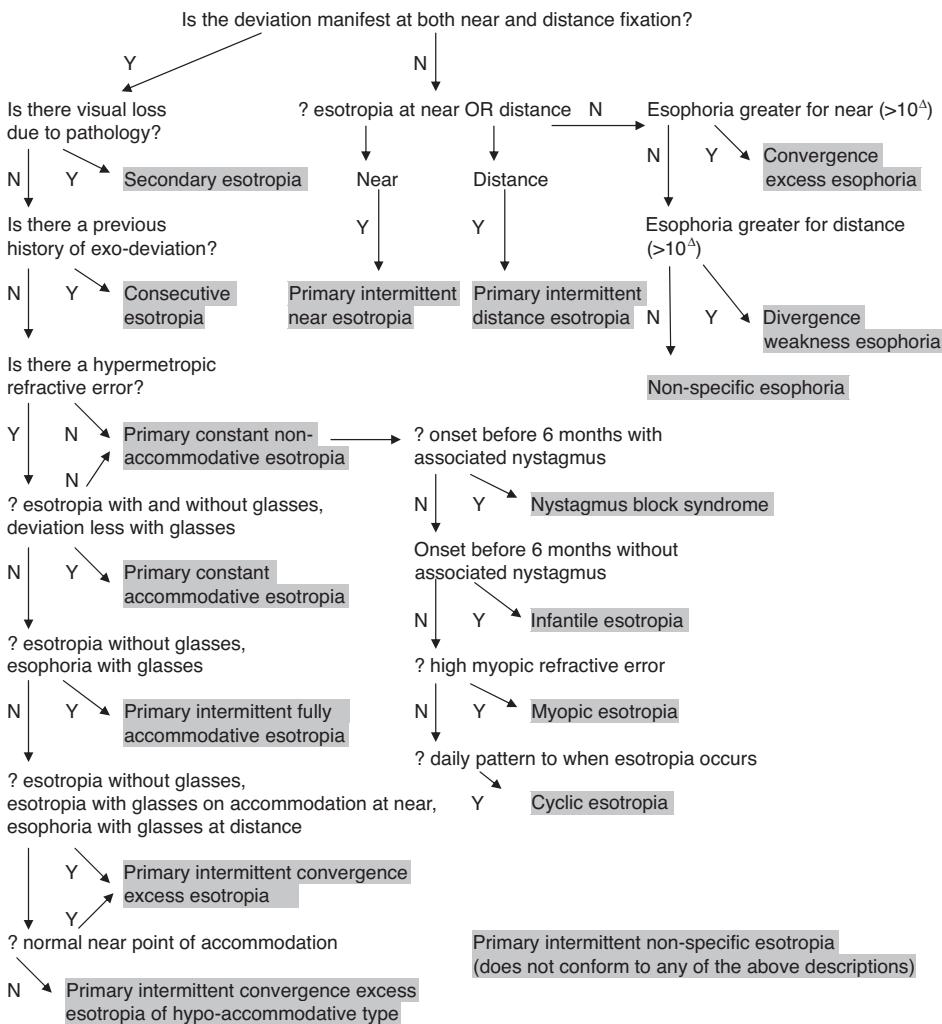


Chart 1 Eso-deviation. (Rowe FJ. Diagnosis of concomitant strabismus. Eye News. 2003; 9: 18.)

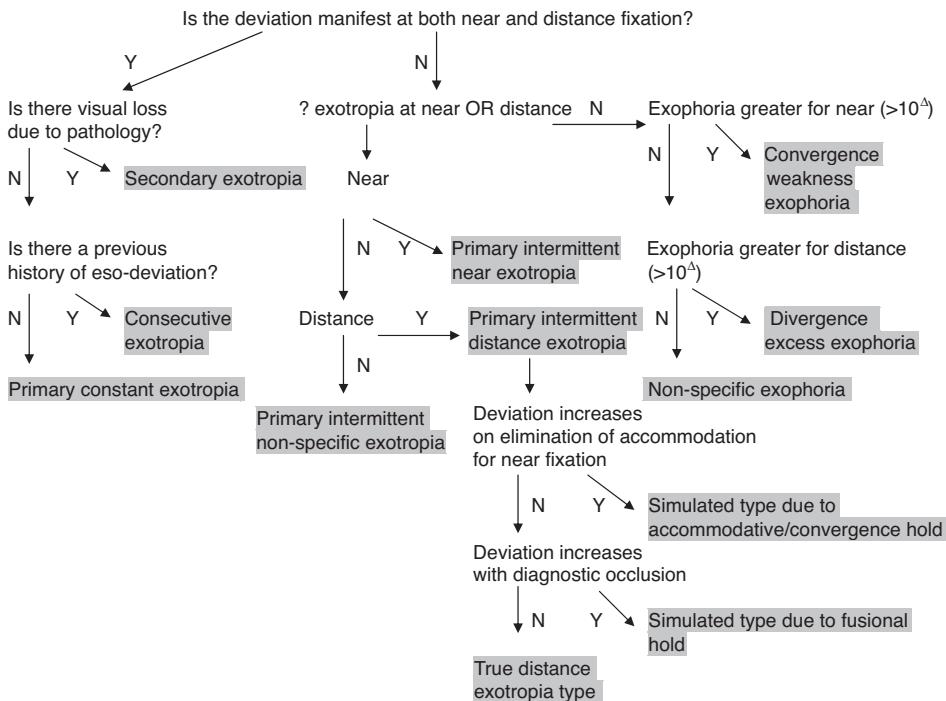


Chart 2 Exo-deviation. (Rowe FJ. Diagnosis of concomitant strabismus. Eye News. 2003; 9: 18.)

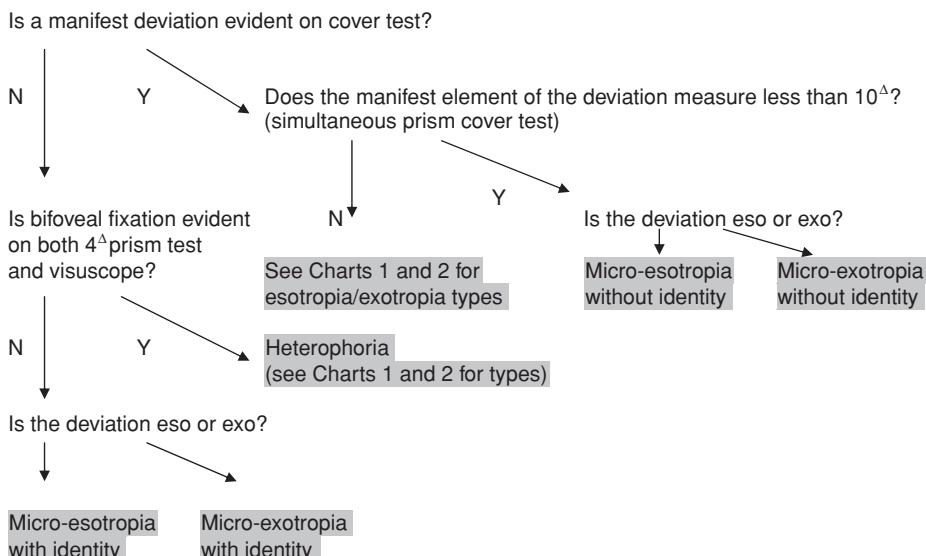


Chart 3 Microtropia. (Rowe FJ. Diagnosis of concomitant strabismus. Eye News. 2003; 9: 18.)

Further reading

- Rowe FJ. Use of Orthoptic investigations in the assessment of incomitant strabismus. Eye News. 2002; 9: 7.
- Rowe FJ. Diagnosis of concomitant strabismus. Eye News. 2003; 9: 18.

Abbreviations of Orthoptic Terms

Abd	Abduction
Accom	Accommodation
AC/A	Accommodative convergence/accommodation ratio
ACS	Alternating convergent strabismus
Add	Adduction
Adj	Adjustable suture
ADS	Alternating divergent strabismus
AHP	Abnormal head posture
Alt	Alternating
Amb	Amblyopia
ARC	Abnormal retinal correspondence
A ‘eso’	A pattern esotropia/esophoria
A ‘exo’	A pattern exotropia/exophoria
bd/bid	Twice daily (bis die/bis in die)
Bag. gls.	Bagolini glasses
BD	Base-down prism
BE	Both eyes
BEO	Both eyes open
BF	Binocular function
BHHT	Bielschowsky head tilt test
BI	Base-in prism
BO	Base-out prism
BO#	Blow-out fracture
BSV	Binocular single vision
BTXA/BT	Botulinum toxin A
BVA	Binocular visual acuity
BU	Base-up prism
CBA	Controlled binocular acuity
CC	Cardiff cards (vanishing optotypes)
CF	Counting fingers
CH	Case history

CHP	Compensatory head posture
CI	Convergence insufficiency
c/o	Complains of
Conv.	Convergence
CPD	Cycles per degree
CPEO	Chronic progressive external ophthalmoplegia
CR	Corneal reflections
CSM	Central steady maintained (fixation)
CT	Cover test
Cyl	Cylinder
D	Dioptrē
Del. rec.	Delayed recovery
Dep.	Depression
DHD	Dissociated horizontal deviation
Dip	Diplopia
DistET/DET	Distance esotropia
DistXT/DXT	Distance exotropia
Div	Divergence
DS	Dioptrē sphere
DV/DVA	Distance vision
DVD	Dissociated vertical deviation
DVM	Delayed visual maturation
Ecc. fix.	Eccentric fixation
EE	Either eye
Elev.	Elevation
EOM	Extraocular muscle
EP	Esophoria
Eso	Esodeviation
ET	Esotropia
E(T)	Intermittent esotropia
Exo	Exodeviation
FCPL	Forced choice preferential looking
FD2	Frisby Davis 2 stereotest
FDT	Forced duction test
FEE	Fixing either eye
FLE	Fixing left eye
FRE	Fixing right eye
FTTO	Full time total occlusion
Gd. rec.	Good recovery
Gls	Glasses
H/A	Headache
H&T	Hundreds and thousands
HoP	Hypophoria
Hot	Hypotropia
HP	Hyperphoria

HT	Hypertropia
HM	Hand movements
INO	Internuclear ophthalmoplegia
IO	Inferior oblique
IPD	Interpupillary distance
IR	Inferior rectus
ISQ	In status quo (in the former state)
KP	Kay's pictures (single optotypes)
L	Left
LCS	Left convergent strabismus
LDS	Left divergent strabismus
LE	Left eye
LN	Latent nystagmus
LogMAR	Logarithm of the minimal angle of resolution
LP	Light perception
LPS	Levator palpebrae superioris
LR	Lateral rectus
L/R	Left hyper/right hypodeviation
LVA	Low vision aid
LVA/LV	Low visual acuity
MA	Major amblyoscope
MBWNL	Muscle balance within normal limits
MLN	Manifest latent nystagmus
MR	Medial rectus
N	Near test type size/near/nerve
NAD	Nil abnormal detected/no apparent deviation.
N&D	Near and distance
NP	Near point
NPA	Near point of accommodation
NPC	Near point of convergence
NPL	No perception of light
Nr	Near
NRC	Normal retinal correspondence
o/a	Overaction
Obj	Objection
Occl	Occlusion
OD	Right eye
OKN	Optokinetic nystagmus
OM	Ocular motility
OS	Left eye
OU	Both eyes
PBD	Prism base down
PBI	Prism base in
PBO	Prism base out
PBU	Prism base up

PCT	Prism cover test
PD	Pupillary distance
PERLA	Pupils equal and reactive to light and accommodation
PF	Palpebral fissure
PFR	Prism fusion range
PH	Pin hole
PL	Preferential looking
PL	Perception of light
PRT	Prism reflection test
PTTO	Part-time total occlusion
RAF	Royal air force (rule)
Rap. rec.	Rapid recovery
RAPD	Relative afferent pupillary defect
RC	Retinal correspondence
RCS	Right convergent strabismus
RDS	Right divergent strabismus
RE	Right eye
RET	Right esotropia
Rec	Recovery
ROP	Retinopathy of prematurity
R/L	Right hyper/left hypo deviation
rr	Rapid recovery
RVA	Right visual acuity
RXT	Right exotropia
Rec/recess	Recession
Res/resect	Resection
SA	Stereoacuity
SFP	Simultaneous foveal perception
SG/SSG	Sheridan Gardiner (singles)
Sl. rec.	Slight recovery
SMP	Simultaneous macular perception
Sn	Snellen's
SO	Superior oblique
SP	Simultaneous perception
Sph	Sphere
sr	Slow recovery
SR	Superior rectus
Subj	Subjective
Supp	Suppression
SV	Stereo vision
tid/td	Three times daily (ter in die/ter die)
u/a	Underaction
VA	Visual acuity
VEP	Visual evoked potential
VER	Visual evoked response

V 'eso'	V pattern esotropia/esophoria
V 'exo'	V pattern exotropia/exophoria
VF	Visual field
VFA	Visual field assessment
VOR	Vestibulo-ocular reflex
X pattern	X pattern
XP	Exophoria
XT	Exotropia
X(T)	Intermittent exotropia
'	Minute of arc
"	Second of arc
Δ	Prism dioptre
	No deviation
	No horizontal deviation
	No vertical deviation
°	Degree
1/3m	One-third metre testing distance
3m	Three metre testing distance
6m	Six metre testing distance
With	With
Without	Without
>	Greater than
<	Less than
1/7	One day
1/52	One week
1/12	One month
Rx	Prescription
Sx	Surgery
#	Fracture

DIAGRAMMATIC ABBREVIATIONS

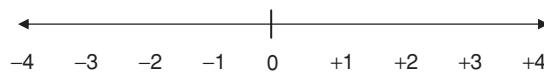
E	Esophoria
RET	Right esotropia
R(E)T	Intermittent right esotropia predominantly controlled
RE(T)	Intermittent right esotropia predominantly manifest
RH	Right hyperphoria
RHT	Right hypertropia
R(H)T	Intermittent right hypertropia predominantly controlled
RH(T)	Intermittent right hypertropia predominantly manifest
Rhyp	Right hypophoria

RhypoT	Right hypotropia
R(Hypo)T	Intermittent right hypotropia predominantly controlled
Rhypo(<u>T</u>)	Intermittent right hypotropia predominantly manifest
RXT	Right exotropia
R(X)T	Intermittent right exotropia predominantly controlled
RX(<u>T</u>)	Intermittent right exotropia predominantly manifest
X	Exophoria

Replace R (right) with L (left) or A (alternating).

Diagrammatic Recording of Ocular Motility

Ocular movements are graded on a 9-point scale of -4 through to $+4$. Zero represents normal full movement. Negative values relating to underactions or limitations and positive values relating to overactions.



-4 represents no movement of the eye(s) past midline (marked).

-3 represents 75% deficit of movement (moderate).

-2 represents 50% deficit of movement (small).

-1 represents 25% deficit of movement (slight).

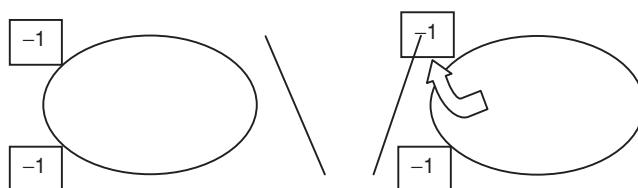
$+$ values represent similar percentages of excess movement.

Hashed lines represent limitations of movement.

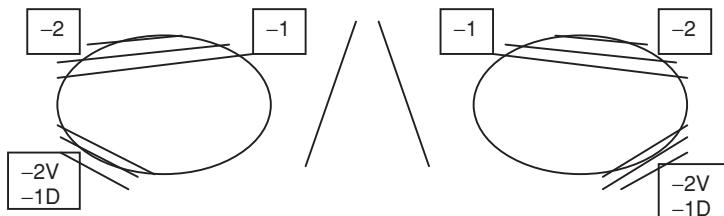
D: Ductions.

V: Versions.

Small superior oblique underaction with small ipsilateral inferior oblique overaction (and updrift), slight contralateral inferior rectus overaction and superior rectus underaction plus V pattern.



Thyroid eye disease case with small restriction of either eye on elevation and depression in abduction with improvement of movement on ductions on downgaze to slight restriction plus A pattern.



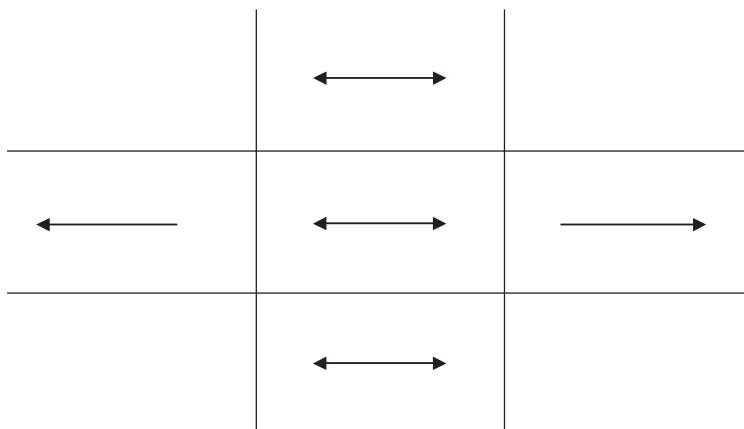
Further reading

O'Flynn EA. Strabismus documentation: an alternative approach. British Orthoptic Journal. 1994; 51: 10–14.

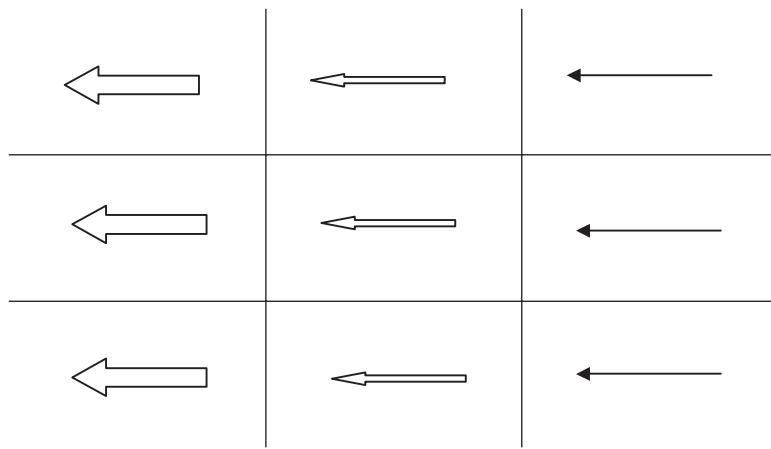
Diagrammatic Recording of Nystagmus

- Right beating jerk nystagmus
- ← Left beating jerk nystagmus
- ↔ Pendular nystagmus
- Increasing amplitude on right gaze
- ←← Increasing amplitude on left gaze
- ↑↑ Increasing amplitude on up gaze
- ↓↓ Increasing amplitude on down gaze
- Increasing frequency on right gaze
- ←←← Increasing frequency on left gaze
- ↑↑↑ Increasing frequency on up gaze
- ↓↓↓ Increasing frequency on down gaze

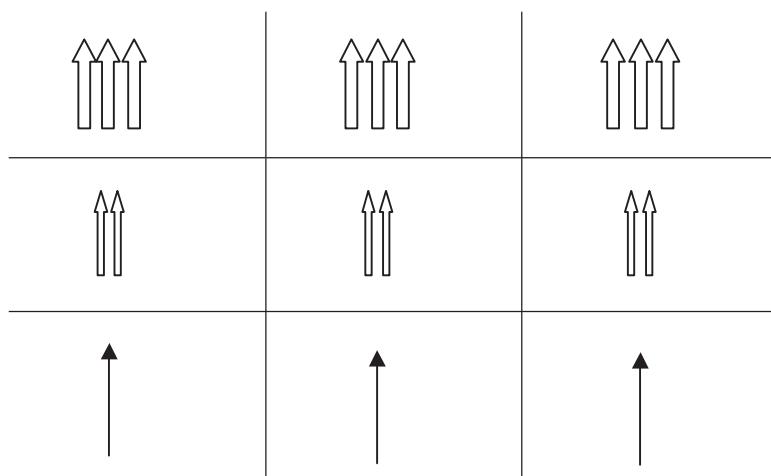
Horizontal pendular nystagmus in upgaze and downgaze becoming horizontal jerk nystagmus on lateral gaze.



Right beating horizontal jerk nystagmus in all positions of gaze, increasing in amplitude on right gaze.



Upbeating nystagmus increasing in amplitude and frequency on upgaze.



Glossary

Abduction

Unocular eye movement where the eye is rotated outwards from the midline.

Abnormal binocular single vision

A form of binocular vision that occurs in the absence of bifoveal fixation, usually with abnormal retinal correspondence in everyday sight.

Abnormal head posture

An abnormal position of the head due to ocular or non-ocular causes. This may consist of:

- chin elevation;
- face turn to the right or left;
- head tilt to the right or left shoulder.

These components may exist together or singly.

Abnormal retinal correspondence

A binocular condition in which there is a change in visual projection such that the fovea of the fixing eye has a common visual direction with an area other than the fovea of the deviating eye. The pairing of all retinal elements is similarly changed. It may occur whichever eye is used for fixation.

Accommodation

The ability of the eye to increase its dioptric power (the convexity of the crystalline lens) in order to obtain a clear image of a near object.

Accommodative convergence/accommodation (AC/A) ratio

Ratio of accommodative convergence in prism dioptres in relation to one dioptre sphere of accommodation.

Accommodative esotropia

The convergent deviation is affected by the state of accommodation and this is a significant factor in the aetiology of the squint.

Accommodative facility

A measure of how fast clarity is restored following a rapid change of focus.

Adduction

Uniocular eye movement to describe rotation inwards towards the midline.

Alexander's law

Relating to nystagmus where gaze in the direction of the fast component increases the frequency and amplitude while gaze in the opposite direction has the reverse effect.

Alternating strabismus

Either eye may be used for fixation.

Amblyopia

A condition of diminished visual form sense that is not the result of any clinically demonstrable anomaly of the visual pathway and which is not relieved by the elimination of any defect that constitutes a dioptric obstacle to the formation of the foveal image.

Amblyopic (amblyogenic) factors

Factors that are thought to cause amblyopia are as follows:

1. Form deprivation at the fovea
2. Abnormal binocular interaction
3. Light deprivation of the entire retina

Amplitude of accommodation

The difference in dioptres between the near and far points of accommodation (between the minimum and maximum amount of accommodation).

Angle kappa

The angle between the mid-pupillary line and the visual axis.

Angle of anomaly

The difference between the objective and subjective angles of deviation.

Aniseikonia

A difference in size and/or shape of the retinal images of the two eyes.

Anisometropia

A difference in refractive error between the two eyes.

Antagonist

Extraocular muscle whose action opposes that of the contracting muscle.

Aphakia

Absence of the crystalline lens from the pupillary line.

Asthenopia

The collective term for various ocular symptoms, including sore and aching eyes, blurred vision and ocular fatigue.

Bell's phenomenon

Elevation of both eyes with divergence when the eyelids are closed.

Bifoveal fixation

Imaging of an object on both foveae at the same time.

Binocular convergence

Convergence associated with binocular single vision.

Binocular diplopia

This results from the presence of a manifest ocular deviation and is the simultaneous appreciation of two separate images caused by the stimulation on non-corresponding points by one object. This separation may be horizontal, vertical or torsional, or any combination.

Binocular function

The ability of the two eyes to coordinate binocularly either in free space, using prisms or instrumentally using a haploscope. It may be normal or abnormal.

Binocular single vision

The ability to use both eyes simultaneously so that each eye contributes to a common single perception and may be bifoveal or monofoveal.

Binocular visual acuity

The maximum visual acuity obtainable while maintaining binocular single vision (as distinct from visual acuity with both eyes open).

Central fixation

The reception of the image of the fixation object by the fovea, the fixation object lying in the principal visual direction.

Cog-wheel movements

Episodic rotations with pursuit movements.

Cogan's lid twitch

Transient eyelid retraction during refixation from down to straight ahead seen in myasthenia gravis.

Cogan's sign

A tonic deviation of the eye occurring on forced eyelid closure and associated with lesions in the cerebral hemispheres.

Collier's sign

Unilateral or bilateral lid retraction due to a lesion in the midbrain.

Coloboma

A fissure or gap in the eyeball or one of its parts, for example iris.

Concomitant strabismus

Strabismus in which the angle of deviation remains the same in all directions of gaze, whichever eye is fixing.

Confusion

The simultaneous appreciation of two superimposed images due to the stimulation of corresponding retinal points by two different images.

Conjugate movements

Movements of the two eyes in the same direction.

Contracture

The inability of a contracted muscle to relax fully.

Contrast sensitivity

The ability of the eye to detect objects of varying contrast that can be tested using sinusoidal gratings of varying spatial frequency and varying luminance intensity.

Convergence

Simultaneous rotation of the eyes inwards.

Convergence accommodation/convergence ratio (CA/C Ratio)

Ratio of convergence initiated accommodation per prism dioptre of convergence.

Crossed fixation

Patient uses right eye to fix in left field and left eye to fix in right field; seen in early onset esotropia, strabismus fixus and sixth nerve palsy. *See also Tripartite Fixation.*

Crowding phenomenon (separation difficulty)

The phenomenon in which a line of letters or symbols of the same size on a test type are identified less easily than single optotypes.

Depression

Rotation of the eye downwards.

Dioptre

A unit of measurement of the power of a lens equal to the inverse of the focal length in metres. A 1-dioptre lens brings parallel light in focus at 1 m.

Diplopia

The simultaneous appreciation of two images of one object.

Disjunctive movements

Movements of the two eyes in which the visual axes do not remain parallel but move in opposite relative directions.

Divergence

Simultaneous rotation of the eyes outwards.

Donder's law

To each position of the line of sight belongs a definite orientation of the horizontal and vertical retinal meridians relative to the coordinates of space irrespective of the route taken.

Duction

Rotary movement of one eye from the primary position.

Eccentric fixation

A uniocular condition in which there is fixation of an object by a point other than the fovea. This point adopts the principal visual direction. The degree of the eccentric fixation is defined by its distance from the fovea in degrees.

Eccentric viewing

A uniocular condition in which there is fixation of an object by a retinal point other than the fovea without change in the principal visual direction.

Ecchymosis

A bruise: an effusion of blood under the skin.

Elevation

Rotation of the eye upwards.

Enophthalmos

Recession of the eyeball into the orbit.

Epicanthus

Skin folds between the medial canthi and the nose causing pseudostrabismus.

Epiphora

An excessive flow of tears.

Epistaxis

Bleeding from the nose.

Esophoria

On dissociation of the two eyes the occluded eye deviates nasally.

Esotropia

One or other eye deviates nasally when both eyes are open.

Exophoria

On dissociation of the two eyes the occluded eye deviates temporally.

Exophthalmos

Protrusion of the eyeball.

Exotropia

One or other eye deviates temporally when both eyes are open.

Faden

A surgical procedure where the muscle is stitched to the globe, as far as possible behind the insertion, to further weaken its action in its field of gaze.

False localisation

The incorrect uniocular subjective localisation of the fixation object.

Far point of accommodation

The furthest point at which an object can be seen clearly.

Field of binocular single vision

The extent to which binocular single vision is maintained by movement of the eyes whilst the head is kept still.

Field of uniocular fixation

The extent to which each eye can maintain foveal fixation whilst the head is kept still.

Fixation disparity

A phenomenon which occurs in binocular single vision in which the image is seen singly despite a slight under or over convergence of the visual axes, provided the disparate retinal points are within Panum's area. It generally increases if BSV is under stress and can be demonstrated instrumentally when fusion occurs for identical features of two targets but any dissimilar features are displaced in the direction of the heterophoria.

Fresnel lenses/prisms

Plastic lenses or prisms that can be adhered temporarily to spectacles.

Functional

1. A deviation that has the ability, at least potentially, for binocular single vision.
2. Patient shows signs or symptoms of a disorder but careful examination fails to reveal any evidence of structural or physiological abnormalities.

Fusional vergence

The range of convergence and divergence of the eyes whilst maintaining clear binocular single vision.

Gait

Manner of walking.

Glissade

A small corrective movement slower than a saccade, used to fixate an object after a hypo- or hypermetric saccade.

Grades of binocular vision

These are the aspects of binocular function that indicate the degree of binocular vision present, that is, simultaneous perception, superimposition, fusion and stereopsis.

Harmonious abnormal retinal correspondence

The angle of anomaly is equal to the objective angle.

Hering's law

A law of ocular motor innervation. Whenever an impulse for the performance of an eye movement is sent out, corresponding muscles of each eye receive equal innervations to contract or relax.

Heteronymous (crossed) diplopia

Binocular diplopia associated with exotropia in which the image of the fixation object is received on the temporal area of the retina of the deviating eye and is projected nasally.

Homonymous (uncrossed) diplopia

Binocular diplopia associated with esotropia in which the image of the fixation object is received on the nasal area of the retina of the deviating eye and is projected temporally.

Horopter

The locus of all points in space that are imaged on corresponding retinal points.

Iatrogenic

A disorder resulting from treatment.

Idiopathic

Without apparent cause.

Incomitant strabismus

Strabismus in which the angle of deviation differs depending upon the direction of gaze or according to which eye is fixing, associated with:

1. defective movement of the eye;
2. asymmetrical accommodative effort.

Infantile

Occurring in the first months of life.

Intermittent strabismus

A manifest deviation that is only present under certain conditions.

Interpupillary distance

The distance between the two pupillary centres. The average for an adult is 65 mm when the eyes are fixing in the distance.

Ipsilateral

Referring to the same side.

Lid lag

A condition occurring in thyroid eye disease where the upper lid does not follow the eye fully when changing fixation from upgaze to downgaze. An area of white sclera will be exposed (von Graefe's sign).

Lid retraction

Raised upper lid due to overaction of Muller's muscle.

Linear optotype

Letters, symbols or pictures presented in evenly spaced rows with separation between the letters equal to the size of the letter.

Linear visual acuity

Visual acuity tested using a line of optotype.

Listing's law

A law elaborating Donder's law of combined actions of the extraocular muscles: each movement of the eye from the primary position to any other position involves a rotation around a single axis lying in the equatorial plane called Listing's plane.

LogMAR

The logarithm of the minimum angle of resolution.

Macropsia

Objects appear larger than their natural size.

Mechanical strabismus

A squint caused by obstruction to movement of the extraocular muscles.

Micropsia

Objects appear smaller than their natural size.

Miosis

Constriction of the pupil.

Monocular diplopia

Double vision attributable to one eye.

Motor fusion

The ability to maintain sensory fusion through a range of vergence that may be horizontal, vertical or cyclovergence.

Muscle sequelae

Sequence of extraocular muscle adaptation following muscle weakness or limitation. Relates to Hering's and Sherrington's laws of innervation.

Mydriasis

Dilation of the pupil.

Myectomy

Removal of a portion of a muscle.

Myopathy

A primary disease of the muscle, causing it to underact.

Myositis

Inflammation of a muscle.

Myotomy

Incision into the muscle.

Near point of accommodation

The nearest point at which an object can be seen clearly.

Near point of convergence

The nearest point at which convergence can be achieved.

Near triad

Association between convergence, accommodation and pupil reflexes.

Negative fusional range/vergence

Motor fusion maintained with base-in prisms or with the eye diverging.

Negative relative convergence

Exerting less convergence in association with the required accommodation.

Neuropathy

A disorder affecting the structure and function of the nervous system.

Normal retinal correspondence

A binocular condition in which the fovea and areas on the nasal and temporal side of one retina correspond to and have a common visual direction with the fovea and temporal and nasal areas of the retina of the other eye.

Null position of nystagmus

Position of the eyes in which the amplitude of nystagmus is least.

Objective

Responses observed by the examiner.

Objective angle

The angle of misalignment of the visual axes as measured by the observer.

Occluder

A device for obscuring the vision of one eye, either totally or partially, so as to ensure the use of the other eye.

Occlusion

The reduction or elimination of retinal image in order to prevent or reduce visual stimulation.

Ocular adnexa

Term applied to eyelids, lacrimal glands, accessory nasal sinuses, etc.

Optokinetic nystagmus

Normal oscillatory eye movements that occur with movement of the visual environment.

Optotype

Letter, symbol or picture used to test visual acuity.

Oscillopsia

An illusion of oscillatory movement of the environment experienced in patients with nystagmus.

Overaction

Excessive action of a muscle caused by increased innervation as a consequence of palsy or limitation to the ipsilateral antagonist or contralateral synergist.

Palpebral fissure

The distance/gap between the upper and lower lids.

Palsy

Partial or complete loss of function of a muscle.

Panoramic vision

The awareness of an enlarged field of vision due to the separation of visual fields occurring in exotropia.

Panum's area

The area surrounding corresponding retinal points within which disparity of correspondence may occur whilst maintaining binocular single vision.

Panum's space

A band around the horopter in space within which object points give rise to binocular single vision.

Paradoxical diplopia

Pathological binocular diplopia in which heteronymous (crossed) diplopia occurs in esotropia or homonymous (uncrossed) diplopia occurs in exotropia.

Paralysis

Complete inability of a muscle to act.

Paresis

Partial paralysis or defective action in a muscle.

Penalisation

The treatment of amblyopia and/or eccentric fixation by optical reduction of form vision of the non-amblyopic eye at one or all fixation distances. The effect may be achieved by the alteration of the spectacle correction and/or use of a cycloplegic drug.

Photophobia

An intolerance of light.

Physiological diplopia

A type of diplopia that exists in the presence of binocular single vision. It consists of the appreciation that a near object appears double when a distance object is fixated (heteronymous or crossed diplopia) and a distant object appears double when a near object is fixated (homonymous or uncrossed diplopia).

Positive fusional range/vergence

Motor fusion maintained with base-out prisms or with the eyes converging.

Positive relative convergence

Exerting more convergence in association with the required accommodation.

Primary angle of deviation

The deviation when fixing with the unaffected eye in paralytic incomitant strabismus.

Principal visual direction

The line of projection in subjective space of images stimulating the fovea or eccentric point used for fixation.

Prism

A wedge-shaped refracting media that deflects traversing rays of light towards the base.

Prism dioptre

A unit of measurement of the strength of a prism. A prism of 1 dioptre displaces the image of the object through 1 cm for each metre of distance between the object and the prism.

Projection

The subjective interpretation of the direction of an object.

Proptosis

Protrusion of one or both eyeballs.

Recession

A surgical procedure that weakens the action of the muscle by moving the insertion towards the origin.

Resection

A surgical procedure that strengthens the action of the muscle by removing a portion close to the insertion. The muscle is then reattached to the original insertion.

Retinal correspondence

Retinal areas of each eye have the same visual direction during binocular vision.

Retinal rivalry

Occurs in binocular vision when non-fusible images are presented to corresponding retinal points. The images are alternately suppressed, ‘shimmer’ or form a changing mosaics.

Saccade

Rapid eye movement to obtain fixation on an object.

Sagittalisation

A surgical procedure where the insertion of the oblique muscle is moved more anteriorly that enhances the torsional action.

Scotoma

An area of partial or complete blindness surrounded by normal or relatively normal visual field.

Secondary angle of deviation

The deviation when fixing with the affected eye in paralytic incomitant strabismus.

Sensory fusion

The ability to perceive two similar images, one formed on each retina, and interpret them as one.

Sherrington's law

A law of reciprocal innervation; whenever an agonist receives an impulse to contract, an equivalent inhibitory impulse is sent to its antagonist which relaxes.

Simultaneous perception

The ability to perceive simultaneously two images, one formed on each retina.

Smooth pursuit movement

Movement of the eyes to maintain fixation on a moving object.

Stereoacuity

An angular measurement of the minimum resolvable binocular disparity necessary for the appreciation of stereopsis.

Stereoscopic vision

The perception of relative depth of objects on the basis of binocular disparity.

Strabismus

A manifest or latent ocular deviation.

Subjective

Response from the patient or subject.

Subjective angle

The angle between the visual directions of the retinal element in each eye receiving images of the fixation object. It is shown by the subjective separation of diplopia in heterotropia or dissociated heterophoria.

Superimposition

The simultaneous perception of two images formed on corresponding areas, with the projection of these images to the same position in space. This may occur whether the correspondence is normal or abnormal. If fusion is absent, two similar images are seen as separate but superimposed and no fusion range is demonstrable.

Suppression

The mental inhibition of visual sensations on one eye in favour of the other eye, when both eyes are open. This may occur in binocular single vision and in manifest strabismus. It may vary in area and density. A defined area is known as suppression scotoma.

Synergists

Contralateral muscles that normally work together.

Tenectomy

A surgical procedure that removes a portion of the tendon, thereby weakening its action (usually the superior oblique).

Tenotomy

A surgical procedure that cuts into the tendon, thereby weakening its action (usually the superior oblique).

Transposition

A surgical procedure that moves the insertion of the muscle to alter its action.

Tuck (plication)

A surgical procedure that shortens the tendon and thereby strengthens its action (usually performed on superior oblique).

Underaction

Reduced ocular rotation which improves on testing ductions, often associated with neurogenic palsies.

Unharmonious abnormal retinal correspondence

The angle of anomaly is less than the angle of deviation.

Version

Movement of the two eyes in the same direction.

Visual direction

The line of projection in subjective space associated with a given retinal point.

Visual Impairment

Any loss of abnormality of visual function.

Yoke muscle

See Synergists.

Case Reports

Concomitant strabismus

Case report 1

Refraction

There was no significant refractive error.

Fundus and media

Normal.

Case history

A child was referred by his general practitioner at the age of 6 months with a history of a convergent deviation from the age of 1 month. There was no significant family history.

Cover test

Marked angle-alternating esotropia.

Ocular motility

The child used cross-fixation on lateral gaze but ocular movements were full on testing.

Angle of deviation

60 prism dioptres.

Diagnosis

A diagnosis of infantile esotropia was made.

Management

The child was reviewed over the following 8 months and surgery of bilateral medial rectus recessions was performed at the age of 14 months. Post-operatively, the eyes appeared aligned although it was not possible to demonstrate evidence of binocular function due to the young age.

Orthoptic examination at the age of 4 years demonstrated a minimal left microtropia with good binocular function.

Case report 2

Case history

A female aged 7 years attended for a follow-up appointment at the orthoptic clinic. An intermittent alternating convergent deviation had been noted from the age of 4 years with no symptoms of diplopia.

Refraction

She had a hypermetropic refractive error and was currently wearing +3.50 DS before either eye.

Visual acuity

Equal and normal visual acuities of 6/6 right and left.

Cover test

Slight angle esophoria with good recovery with the spectacles and a moderate angle right esotropia without the spectacles.

<i>Binocular function</i>	Normal with the spectacles with a prism fusion range of 45 prism dioptres convergent range and 12 prism dioptres divergent range. Binocular visual acuity was to the level of visual acuity with the spectacles and there was bifoveal fixation evident with the 4 prism dioptre test.
<i>Diagnosis</i>	A diagnosis of intermittent accommodative esotropia of the fully accommodative type was made.
<i>Management</i>	The spectacles were reviewed on a yearly basis and no further management was required.

Case report 3

<i>Refraction</i>	Retinoscopy revealed a hypermetropic refractive error and spectacles of +2.0 DS right and left were ordered.
<i>Case history</i>	A 4-year-old female was referred by her general practitioner with a history of intermittent convergent squint over the past year. There was a history of hypermetropic refractive error in the immediate family.
<i>Visual acuity</i>	Equal.
<i>Cover test</i>	Small angle esophoria to a light target for near fixation becoming a moderate angle right esotropia to a detailed target. There was a slight angle esophoria in the distance. Without the spectacles, there was a moderate angle right esotropia on near fixation and a moderate angle esophoria for distance that was easily dissociated to a right esotropia.
<i>Binocular function</i>	Poor responses at near fixation. However, a normal motor fusion range and stereopsis was demonstrable using the synoptophore.
<i>Angle of deviation</i>	The angle of deviation measured 40 prism dioptres for near fixation and 14 prism dioptres for distance.
<i>+3.0 DS lenses</i>	The deviation was again measured with additional +3.0 DS lenses for near fixation and the angle of deviation decreased to 16 prism dioptres.
<i>AC/A ratio</i>	An AC/A ratio of 8:1 was calculated by the gradient method.
<i>Diagnosis</i>	A diagnosis of intermittent accommodative exotropia of the convergence excess type was diagnosed.
<i>Management</i>	The angle of deviation was found to increase on testing at follow-up visits and binocular function deteriorated. Surgery was planned and bilateral medial rectus recessions were done at the age of 5 years. Post-operatively, there was a small residual manifest angle of deviation on near fixation with a latent deviation in the distance. This persisted and 2 months post-operatively, the child was given miotic drops. The manifest deviation was controlled for near fixation with good binocular function while using these drops. After 2 months of full control of the deviation, the drops were gradually withdrawn over a further 2-month period. The child has retained good binocular function and a small angle esophoria is present on cover test.

Case report 4*Refraction**Case history**Visual acuity**Cover test**Ocular motility**Convergence**Binocular function**Angle of deviation**+3.0 DS lenses**AC/A ratio**Diagnosis**Management*

There was no significant refractive error on retinoscopy.

A 5-year-old boy was referred with a history of sore eyes when watching TV. His father also noted that the eyes appeared to lack coordination at times.

Equal in either eye.

Small angle exophoria was present for near fixation with delayed recovery. On distance fixation, a moderate angle exophoria compensated easily to a left exotropia. This was more evident on far distance fixation.

Ocular motility revealed a V exo pattern with underactions of the superior rectus of either eye.

Binocular convergence was reduced at 14 cm.

Reduced with a low convergent motor fusion range of 18 prism dioptres.

The angle of deviation measured 12 prism dioptres for near fixation and 30 prism dioptres for distance.

The angle was measured with additional +3.0 DS lenses and noted to be 12 prism dioptres.

An AC/A ratio of 6:1 was calculated using the gradient method.

A diagnosis of intermittent exotropia of the simulated distance exotropia type was made.

Although young, the child was very cooperative and convergence exercises were commenced to improve his level of binocular convergence. Pen convergence exercises were advised initially and after 3 weeks, his convergence level had improved to 8 cm with improvement in his convergent motor fusion range also. He continued with pen convergence exercises and was given a dotcard to use as well. Binocular convergence improved to the normal level of 6 cm. At follow-up, exercises were stopped.

He maintained good binocular function with control of the latent deviation for near fixation. As the deviation was not cosmetically unacceptable for distance, further management was not planned and the child remained under observation.

Case report 5*Case history**Visual acuity**Cover test**Angle of deviation*

A 21-year-old male presented with a cosmetically unacceptable left exotropia. There was a history of a penetrating injury to the left eye at the age of 7 years. A traumatic cataract was removed 6-months later and an intraocular lens implanted. He also had left corneal scarring related to the penetrating injury.

6/6 in the right eye and perception of light in the left eye.

Moderate angle left exotropia by corneal reflections with poor left fixation.

Estimated to be 35 prism dioptres by corneal reflections using the prism reflection test.

<i>Suppression</i>	A post-operative diplopia test elicited no diplopia responses at the corrected angle or when under or over corrected by prisms.
<i>Diagnosis</i>	A diagnosis of secondary exotropia was made.
<i>Management</i>	Surgery was planned for a cosmetic result with the aim to leave the left eye in a slight convergent position.

Case report 6

<i>Case history</i>	A 3-year-old girl presented with a 1-year history of intermittent esotropia. The parents had noted an alternate day tendency to squint and had documented this over a 6-week period.
<i>Visual acuity</i>	Equal in either eye.
<i>Cover test</i>	Marked left esotropia at near and distance fixation.
<i>Binocular function</i>	Absent with suppression responses on testing in free space and on the synoptophore.
<i>Angle of deviation</i>	50 prism dioptres at near and distant fixation.
<i>Observation</i>	In view of the parents' observations of the frequency of the deviation, appointments were arranged for consecutive day assessments. On day 1, the marked left esotropia was evident. On day 2, the child was orthophoric with excellent binocular function. A diagnosis of cyclic esotropia was made.
<i>Diagnosis</i>	A squint chart was documented daily over the follow-up period that showed a break in the pattern with the squint more frequent.
<i>Management</i>	A left medial rectus recession and lateral rectus resection procedure was performed at this stage and post-operatively a minimal esophoria with excellent binocular function was demonstrated. This latent deviation has remained well controlled over a 4-year follow-up period with no return of the cyclic pattern.

Case report 7

<i>Refraction</i>	Spectacles of +1.0 DS for the right eye and +3.75 DS for the left eye.
<i>Case history</i>	A female aged 6 years was referred by her optician, having failed a school vision test. There had been no previous ocular history and no significant family history.
<i>Visual acuity</i>	6/5 in the right eye and 6/36 in the left eye.
<i>Cover test</i>	A minimal left esotropia was noted that increased slightly on dissociated but recovered to the minimal manifest angle.
<i>Binocular function</i>	Demonstrable with a motor fusion range of 30 prism dioptres convergent range and 12 prism dioptres divergent range. Stereopsis was reduced at 400 seconds of arc. Fixation was central and steady in the right eye but was unsteady and nasal to foveal fixation in the left eye.

<i>Angle of deviation</i>	12 prism dioptres by prism cover test and 6 prism dioptres by simultaneous cover test.
<i>Diagnosis</i>	A diagnosis of microesotropia without identity was made.
<i>Management</i>	Occlusion therapy was commenced with occlusion for a minimum of 6 hours daily initially. The visual acuity improved over the follow-up period but did not improve beyond the 6/9 level. Occlusion was discontinued at this stage. No further management was required.

Case report 8*Case history**Visual acuity**Cover test**Binocular function**Diagnosis**Management*

A 5-year-old boy was referred having moved into the area. He had previously been under the care of another hospital. Treatment had included one operation for a convergent deviation that had been present from infancy. This had been successful in aligning the eyes. Visual acuity had been found to be slightly reduced and the child had had 1 year of part-time occlusion treatment.

6/9 in the right eye and 6/6 in the left eye.

Slight right exotropia with dissociated vertical deviation and latent nystagmus.

Suppression responses were obtained.

A diagnosis of consecutive exotropia with dissociated vertical deviation and latent nystagmus was made.

The strabismus was cosmetically good and no further management was indicated.

Case report 9*Case history**Refraction**Visual acuity**Cover test**Ocular motility**Convergence**Accommodation**Binocular function**Diagnosis*

A 15-year-old girl was referred by her general practitioner with general asthenopic symptoms of intermittent headache, blurred vision and jumbling of print when reading. Her general health was good and there was no significant family history.

She had a mild myopic refractive error and had been wearing spectacles for the past 9 months.

Equal visual acuities in either eye.

A small angle exophoria was noted at near fixation and a minimal exophoria for distance.

Full.

Reduced to 25 cm and fatigue was noted on repeated testing.

Slightly defective for the patient's age

Reduced convergent range of the prism fusion range.

A diagnosis of exophoria of the convergence weakness type and convergence insufficiency was made.

Management She was given pen convergence exercises to practice daily and reviewed after 2 weeks at which time the level of binocular convergence had improved to 12 cm. She was then given a dot card in addition to the pen convergence exercises. The level of binocular convergence continued to improve and cat stereograms were given. At the next follow-up visit, binocular convergence was to a normal level of 6 cm. She was given detailed stereograms at this stage. Binocular convergence remained at 6 cm and was well maintained at this level. The convergence range of the prism fusion range had also improved to normal and the patient was asymptomatic. Exercises were discontinued and at follow-up 2 months later, she remained asymptomatic with a normal orthoptic assessment, and was discharged.

Incomitant strabismus

Case report 10

Case history

A 58-year-old male presented with a sudden onset of horizontal diplopia 2 days previously. The diplopia was worse on right gaze and there were no other associated symptoms. He had a history of diabetes for the past 15 years.

Cover test

Right esotropia for near fixation that increased in angle at distance. With a small abnormal head posture of face turn to the right, the deviation was controlled for near fixation but readily decompensated in the distance.

Ocular motility

Small right lateral rectus (VI nerve) palsy.

Angle of deviation

18 prism dioptres at near fixation and 25 prism dioptres at distance.

Binocular function

Diplopia could be joined with 16 prism dioptres for both fixation distances.

General investigation

Neurological assessment including a CT brain scan was normal. However, the diabetes was found to be poorly controlled and the patient's treatment was altered.

Diagnosis

A diagnosis of right sixth nerve palsy due to a probable microvascular episode was made.

Management

A 15 prism dioptre Fresnel lens was fitted to the patient's spectacles.

The lateral rectus palsy gradually improved over the following 4 months. During this time, the Fresnel prism was reduced in strength and eventually discarded.

Case report 11

Case history

A 26-year-old male was seen on the neurology ward. He had been involved in a road traffic accident and had suffered a closed head injury.

<i>Cover test</i>	Complete left ptosis with no lid crease. The patient did not complain of diplopia as the ptosis covered the visual axis. On lifting the lid, the left eye was in a depressed and divergent position. The pupil was semi-dilated.
<i>Ocular motility</i>	Complete left third nerve palsy with limited elevation, adduction and depression of the left eye.
<i>Diagnosis</i>	A diagnosis of traumatic third nerve palsy was made.
<i>Management</i>	An outpatient appointment was arranged for a 3 month follow-up visit. At that time, the ocular movements were seen to be recovering. At his 6-month follow-up, there was a slight limitation of adduction with a residual left exotropia. Elevation and depression of the left eye had recovered. The left exotropic angle of deviation remained static over a further 9-month period and surgery of a left lateral rectus recession was performed. Thereafter, the patient regained binocular single vision in the primary position.

Case report 12

Case history

A 42-year-old female was referred by her general practitioner with a 6-month history of intermittent diplopia and grittiness of her eyes. She was aware of a puffy appearance around her eyes. She had had a thyroidectomy 3 years previously and was currently on thyroxine and clinically euthyroid.

Visual acuity

Reduced at 6/18 in the right eye and 6/12 in the left eye.

Cover test

A slight exophoria with a small right hyperphoria was seen.

Ocular motility

Slight limitation of either eye on abduction. There was a moderate limitation of either eye on elevation with the left eye slightly more limited than the right. The right eye was slightly limited on depression, maximum in an abducted position. Bilateral lid retraction and proptosis was noted.

Binocular function

A good motor fusion range was demonstrable including an extended vertical fusion range consistent with a gradual increase in the vertical deviation.

Hess chart

Confirmed restricted ocular movements.

General investigation

An MRI orbital scan demonstrated enlarged extraocular muscles particularly near the orbital apex. An MRI stir sequence showed increased signal from the extraocular muscles confirming active thyroid eye disease.

Diagnosis

A diagnosis of thyroid eye disease was made. In view of the enlarged extraocular muscles at the orbital apex and the reduced visual acuity, optic compression was diagnosed.

Management

The patient was admitted and commenced on pulsed IV methylprednisolone for immediate medical decompression. She then had a course of oral prednisolone and azathioprine. Visual acuity

was monitored by automated visual field assessment and improved to normal acuities of 6/6. The ocular motility limitations were also noted to improve and MRI demonstrated a reduction of the extraocular muscle bulk. The plan was to reduce the medical treatment very gradually.

The patient did not require further treatment for her ocular motility limitations as she was only aware of intermittent diplopia on extreme elevation and she avoided looking in this position of gaze. She had a good field of binocular single vision in the primary and reading positions.

Case report 13

Case history

A 3-year-old child was referred for a second opinion regarding her esotropia and marked abnormal head posture. Her parents were concerned about the degree of head turn and the cosmesis. Equal.

Visual acuity

The head posture was of a marked face turn to the left

Abnormal head posture

With the head posture, there was a slight esophoria. Without the head posture, cover testing revealed a marked left esotropia with some left enophthalmos.

Cover test

Marked limitation of left abduction with widening of the palpebral fissure and slight limitation of left adduction with moderate retraction of the globe and narrowing of the palpebral fissure. The left eye was seen to upshoot on elevation in adduction. There was slight limitation of right abduction with widening of the palpebral fissure but no obvious limitation or retraction on right adduction.

Ocular motility

With the head posture, there was demonstrable binocular function.

Binocular function

A diagnosis of Duane's retraction syndrome was made.

Diagnosis

The child was reviewed during the following 6 months. The binocular function while adopting the abnormal head posture deteriorated and the latent deviation was noted to decompensate at times. In view of this decompensation and the very marked abnormal head posture, it was decided to list the child for surgery. Bilateral medial rectus recessions and a left lateral rectus recession were performed to reduce the convergent angle of deviation and the globe retraction.

Management

Post-operatively, a small abnormal head posture was still adopted but to a much less degree. The convergent deviation was well controlled with the abnormal head posture with good binocular function and the manifest deviation without the abnormal head posture was reduced in measurement.

Case report 14

<i>Case history</i>	A 74-year-old male presented with a 3-day history of sudden onset vertical diplopia. This was worse when reading. He had had a mild left cerebral vascular accident (CVA) 1 week previously resulting in hand weakness that later resolved.
<i>Visual acuity</i>	Equal.
<i>Cover test</i>	Small right hypertropia with a slight right exotropia on near fixation with diplopia. At distance, there was a slight right hypertropia with diplopia.
<i>Ocular motility</i>	Small underaction of the right on depression in adduction and an overaction of the right eye on elevation in adduction consistent with a right superior oblique (IV nerve) palsy with secondary inferior oblique overaction.
<i>Angle of deviation</i>	5 prism dioptres R/L at near fixation and 3 prism dioptres R/L at distant fixation.
<i>Hess chart</i>	Confirmation of ocular motility findings.
<i>Diagnosis</i>	A diagnosis of fourth nerve palsy due to cerebrovascular accident was made.
<i>Management</i>	The patient was mainly troubled by the diplopia in the near position and a 5 dioptrre Fresnel prism was placed on his reading spectacles that joined the diplopia comfortably. The prism was reduced to a 3 dioptrre strength at subsequent follow-up. As the deviation did not recover any further, this prism was incorporated into the spectacle correction 8 months later.

Case report 15

<i>Case history</i>	A 53-year-old male presented with diplopia on upgaze. He had been assaulted 2 weeks previously resulting in a facial injury. A CT scan showed a right orbital floor fracture with entrapment of tissue.
<i>Abnormal head posture</i>	Small abnormal head posture of chin elevation.
<i>Cover test</i>	Minimal esophoria with slight right hypophoria with this head posture. Without the head posture, the deviation became a manifest left hypertropia with diplopia.
<i>Ocular motility</i>	Slight limitation of the right eye on abduction and adduction and a moderate limitation of the right eye on all elevated positions. There was slight enophthalmos with some tingling over his upper lip.
<i>Management</i>	The patient was reviewed by the maxillofacial team and underwent orbital surgery that freed the entrapped tissue and a Silastic plate was inserted over the fracture site. Post-operatively, the patient still had a small limitation of the right eye on elevation with diplopia. However, he was not bothered by this diplopia and was happy to continue using a small abnormal head posture. No further management was indicated.

Case report 16

<i>Case history</i>	A 3-year-old child was seen in community vision screening and was referred with limited elevation of the right eye.
<i>Refraction</i>	The child was uncooperative on attempts to check refraction and his fundus and media. Therefore, he was admitted for an examination under anaesthetic. There was no significant refractive error and his fundus and media were normal and clear.
<i>Visual acuity</i>	Equal in either eye.
<i>Cover test</i>	Slight exophoria with a minimal right hypophoria.
<i>Ocular motility</i>	Moderate limitation of right elevation that was marked in adduction. On left gaze, a downdrift of the right eye was noted. Motility of either eye was normal on depression.
<i>Binocular function</i>	Excellent with a normal motor fusion range.
<i>Forced duction test</i>	Performed under anaesthetic and positive on elevation in adduction.
<i>Diagnosis</i>	A diagnosis of right Brown's syndrome.

Case report 17

<i>Case history</i>	An 11-year-old child was seen in the ophthalmology casualty clinic. She complained of intermittent blurred vision and diplopia. She had been seen by an optician regarding the blurred vision and was now wearing spectacles with a mild myopic prescription.
<i>Refraction</i>	A cycloplegic refraction revealed a slight hypermetropic refractive error that was not significant. The myopic spectacles were discarded.
<i>Fundus and media</i>	Normal.
<i>Visual acuity</i>	Reduced in both eyes but improved with additional minus lenses.
<i>Cover test</i>	Small esophoria was demonstrated. This increased on dissociation and intermittently became a marked alternating esotropia with diplopia. The pupils were noted to be constricted with the manifest deviation.
<i>Diagnosis</i>	A diagnosis of accommodative convergence spasm was made.
<i>Management</i>	An MRI brain scan was arranged to exclude any organic cause for the condition. This scan was normal in appearance. The child was commenced on atropine drops 1% daily and given +3.0 DS spectacles for close work. In the interim, she was referred to a child psychologist. On questioning, a history of bullying at school was revealed. The school was contacted and this problem addressed. The child was weaned off the atropine over a period of 4 months and has remained asymptomatic.

Case report 18

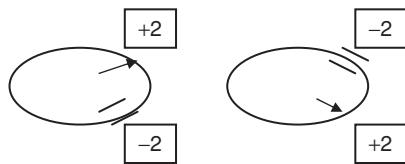
<i>Case history</i>	A 45-year-old male presented to ophthalmology casualty complaining of difficulties reading and headaches.
<i>Investigation</i>	Orthoptic assessment was normal with full ocular movements and a well-compensated exophoria.
<i>Observation</i>	In view of the symptoms, a 1-week follow-up appointment was arranged.
<i>Ocular motility</i>	Downgaze palsy with convergence retraction nystagmus on elevation.
<i>General investigation</i>	A CT brain scan revealed increased signal in the upper midbrain with scattered signal areas in the cortex.
<i>Diagnosis</i>	A diagnosis of disseminating disease was made causing Parinaud's (dorsal midbrain) syndrome.
<i>Management</i>	There was complete recovery of the ocular motility signs during the following 3 months. No treatment other than observation was required.

Case report 19

<i>Case history</i>	A 42-year-old female presented with a 10-day history of right eye pain and diplopia on right gaze that was both horizontal and vertical. She had a medical history of Parkinson's syndrome and medication included antidepressants and sedatives.
<i>Cover test</i>	Slight exophoria with good recovery.
<i>Ocular motility</i>	Slight limitation of the right eye on adduction and moderate limitation of either eye on elevation consistent with an upgaze palsy. There was bilateral ptosis. Saccadic movements were tested that showed defective horizontal saccades and absent saccades on upgaze. Vertical saccades were full on depression.
<i>Convergence</i>	Binocular convergence was reduced to 25 cm.
<i>Diagnosis</i>	A diagnosis of supranuclear ocular motility deficit due to Parkinson's disease was made.

Case report 20

<i>Case history</i>	A 32-year-old male presented complaining of a gradual onset of intermittent blurring of vision and oblique diplopia (combined horizontal and vertical separation of images) for the past year. No other ocular symptoms. General health was good with no medication taken. Previous history of road-traffic accident 10 years ago with concussion. New job in the past year with increasing visual display unit work.
<i>Abnormal head posture</i>	Small tilt and turn to left with chin depression.
<i>Cover test</i>	Slight right hyperphoria and esophoria with abnormal head posture. Angle increases with fair recovery without abnormal head posture, with near angle greater than distance fixation.

Ocular motility*Convergence*

Normal.

*Binocular function**Angle of deviation*

Normal values with abnormal head posture.

R/2 prism dioptres and eso 4 prism dioptres at near fixation, R/1 prism dioptres and eso 4 prism dioptres at distance fixation with abnormal head posture. R/5 prism dioptres and eso 4 prism dioptres at near fixation, R/4 prism dioptres and eso 4 prism dioptres at distance fixation without abnormal head posture and fixing either eye.

Synoptophore

R/3 ^Δ , +3°		R/7 ^Δ , +2°
	R/4 ^Δ , +2°, 1° excyclo	
R/3 ^Δ , +2°, 4° excyclo		R/6 ^Δ , +3°, 2° excyclo

Diagnosis

Diagnosis of right superior oblique paresis as abnormal head posture includes chin depression to move eyes away from position of maximum action of affected muscle, near measurements are greater than distance measurements, symptom of torsion is maximum on downgaze and is excyclo rather than incyclotorsion. Long-standing condition due to concomitant ocular motility and measurements (no difference in primary or secondary angle of deviation), gradual onset of symptoms and not aware of torsion.

Case report 21*Case history*

A 16-year-old female complained of symptom of horizontal diplopia for past 6 months particularly when looking to the right. Not very bothered by diplopia and no other symptoms. General health is good and family history of brother with a squint.

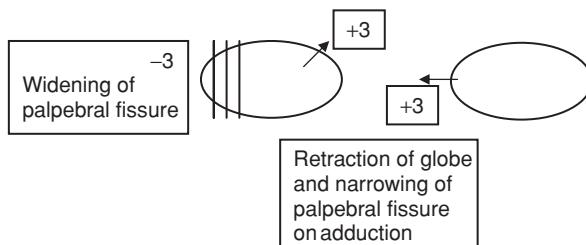
Abnormal head posture

Slight face turn to right.

Cover test

Slight esophoria with abnormal head posture. Slight right esotropia without abnormal head posture greater for distance than near fixation with intermittent diplopia.

Ocular movements



Convergence

Normal.

Binocular Function

Normal values with abnormal head posture. Intermittent suppression/diplopia on right gaze.

Angle of deviation

Eso 6 prism dioptres at near fixation, eso 8 prism dioptres at distance fixation with abnormal head posture. Eso 14 prism dioptres at near fixation, eso 18 prism dioptres at distance fixation without abnormal head posture fixing left eye. Eso 18 prism dioptres at near fixation, eso 25 prism dioptres at distance fixation without abnormal head posture fixing right eye.

Synoptophore



Diagnosis

Diagnosis of Duane's retraction syndrome due to palpebral fissure changes, globe retraction on adduction of the affected eye and upshoot of affected eye on elevation in adduction. Incomitant angle of deviation that increases fixing with the affected eye. Abnormal head posture is only slight and unaware of this. Intermittent symptoms with intermittent suppression/diplopia responses on investigation can be expected in Duane's retraction syndrome. Constant diplopia would be expected more with sixth nerve palsy without suppression responses.

Case report 22

Case history

A 3-year-old boy referred by general practitioner as family had noticed a squint when the child looked up at them. General health is good with no family history of eye problems and a normal birth history.

Cover test

Slight exophoria for near and distance fixation.

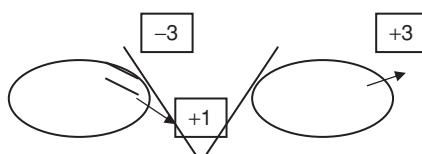
Convergence

Normal

Binocular function

Normal values in primary position.

Ocular movements



Angle of deviation
Synoptophore

L/6 $^{\Delta}$, -4°	L/8 $^{\Delta}$, -3°	L/10 $^{\Delta}$, -3°
	-2°	L/2 $^{\Delta}$, -3°
-2°		-1°

Diagnosis

Diagnosis of Brown's syndrome as muscle sequelae is limited to overaction of the left superior rectus. There is minimal deviation in the primary position with no abnormal head posture and no abnormalities of ocular movement in any depressed position of gaze. A V pattern is present on ocular movements rather than an A pattern.

Case report 23

Case history

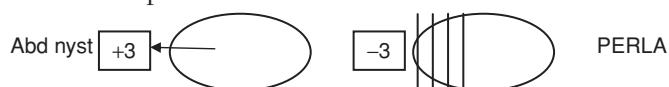
A 72-year-old female presented complaining of constant diplopia. Diplopia still remained after closing one eye. General history was of poorly controlled hypertension and homonymous hemianopia due to a stroke 2 years previously. She had a cataract in her left eye with left monocular diplopia.

6/12 acuity in the right eye and 6/24 in the left eye.

Moderate left exotropia at near fixation with small exophoria at distance fixation.

Absent.

Normal values in primary position when the deviation was corrected with prisms.



Angle of deviation

Diagnosis

Exo 30 prism dioptres on near fixation and exo 14 prism dioptres on distance fixation.

Diagnosis of Left internuclear ophthalmoplegia was made because of the presence of limited adduction with contralateral abducting nystagmus. MRI scan confirmed a small brainstem infarction in the midbrain. Absent convergence indicated the lesion was more rostral in the midbrain. Management was with sector occlusion as the patient had single vision to left gaze.

Case report 24

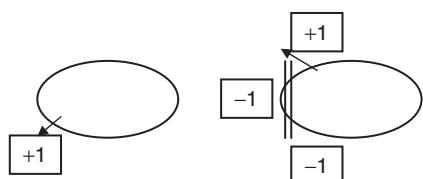
Case history

A 24-year-old male presented complaining of blurred vision, vertical diplopia and twitching of his eye. His general health was good.

Cover test

Small left exotropia and left hypertropia for near and distance fixation.

Ocular movements



Fine L oscillatory movements on dextrodeph.

Angle of deviation

Diagnosis

Exo 25 prism dioptres and L/4 prism dioptres for near and distance fixation.

Diagnosis of Left superior oblique myokymia because of fine oscillatory movements in conjunction with small superior oblique underaction. Symptoms improved when taking carbamazepine.

Case report 25

Case history

A 70-year-old wheelchair-bound male presented complaining of blurred vision and moving images. General health listed relapsing and remitting multiple sclerosis.

Cover test

Horizontal jerk nystagmus was present but no manifest strabismus.

Ocular movements

Horizontal gaze evoked nystagmus was present to right and left gaze with rebound nystagmus on refixation. A rotary element was also seen to the nystagmus. Impaired smooth pursuit gain with occasional cog-wheel saccadic movements were noted along with impaired vestibulo-ocular reflex.

Convergence

Reduced to 18 cm.

Diagnosis

Diagnosis of nystagmus due to multiple sclerosis was made.

The patient was advised that unilateral retrobulbar injection of botulinum toxin might be an option in conjunction with occlusion of the other eye, should the symptoms impact on reading, watching television and other activities of daily living. The patient deferred treatment until such time that symptom might worsen.

Case report 26

Case history

A 58-year-old female was referred from the neurology department having presented with symptoms of nausea and headache, vertical diplopia and visual tilt. The neurology referral documented presence of ataxia.

Cover test

A small right hypotropia was noted at near and distance fixation with symptoms of vertical diplopia and incyclotorsion. Visual tilt was recorded along with fine horizontal jerk nystagmus.

Abnormal head posture

A slight head tilt to the right was adopted to compensate for the visual tilt.

<i>Ocular movements</i>	Slight right ptosis was noted and right pupil miosis. Impaired orbicularis function was demonstrated indicating VII paresis. On blinking, the eyes were seen to deviate to the right side (lateropulsion).
<i>Angle of deviation</i>	L/8 prism dioptres for near and distance fixation.
<i>MRI scan</i>	Lateral medullary infarction was confirmed.
<i>Diagnosis</i>	Diagnosis of Wallenberg's syndrome was made consisting of right-sided skew deviation, ocular tilt reaction, lateropulsion and nystagmus. Diplopia was alleviated with a small base-up prism before the right eye.

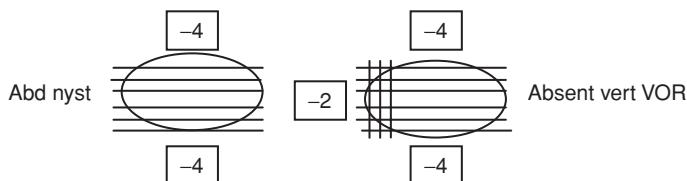
Case report 27*Case history*

A 70-year-old female was referred from the stroke unit when the stroke team noted impaired eye movements. She had mild receptive aphasia and suspected right-sided homonymous hemianopia. The MRI report in the case notes confirmed an upper midbrain infarct affecting the riMLF, interstitial nucleus of Cajal and MLF.

Visual acuity

Acuities of 0.0 logMAR were recorded in either eye using a matching card and the right hemianopia was confirmed.

A small exophoria was present. Diplopia only on right gaze.

*Angle of deviation*

Exo 16 prism dioptres for near and distance fixation.

Diagnosis

Diagnosis of complete vertical gaze palsy impairing both upgaze and downgaze plus left partial internuclear ophthalmoplegia with contralateral abducting nystagmus. Sector occlusion was given on the medial part of the patient's left glasses lens to alleviate diplopia experienced on right gaze. Yoked base-up prisms were placed on the patient's reading glasses to aid with reading as the patient complained of difficulty being able to read due to inability to depress the eyes.

Case report 28*Case history*

A 16-year-old female was referred from the neurosurgical team. She had undergone neurosurgery 6 months previously to remove a craniopharyngioma during which the optic chiasm had been damaged. She was complaining of difficulty with judging distance and intermittent blurred vision.

<i>Visual acuity</i>	Visual acuities of 0.2 (right) and 0.6 (left) logMAR were noted with confirmation of a complete bitemporal hemianopia.
<i>Cover test</i>	At near and distance fixation, a small exophoria was present that decompensated to small exotropia at times with poor recovery.
<i>Ocular movements</i>	Full ocular rotations of either eye.
<i>Convergence</i>	Binocular to 20 cm with break in convergence and diplopia.
<i>Binocular function</i>	Prism fusion range at near fixation was base-out 4 and base-in 2 prism dioptres. No stereopsis could be elicited.
<i>Angle of deviation</i>	Exo 18 prism dioptres for near and distance fixation.
<i>Diagnosis</i>	Diagnosis of hemifield slide was made because of her intermittent decompensation of the exophoria and poor binocular control. This was due to the complete bitemporal hemianopia with loss of normal retinal correspondence. Prisms could not successfully aid control of the deviation and the patient accepted Bangerter foils on her glasses to aid symptoms when reading.

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