Incomitant Strabismus and Principles of Its Management

4

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4.1 Introduction

Differentiation between comitant and incomitant strabismus is an important task for the general ophthalmologist managing strabismus. The paralyses or restrictions underlying incomitant strabismus may imply potentially dangerous disorders that should be treated promptly. Early detection and proper management are necessary to achieve good clinical outcome.

4.2 Basics of Incomitant Strabismus

We initially discuss some concepts related to the understanding of incomitant strabismus. Few of these have already been dealt with in the earlier chapters but are mentioned here to make the understanding clearer.

4.2.1 Comitance and Incomitance

The word "comitant" was originated from the word "concomitant." Concomitant is defined as "accompanying especially in a subordinate or incidental way" in Merriam-Webster Dictionary. It implies that, despite of the strabismic deviation,

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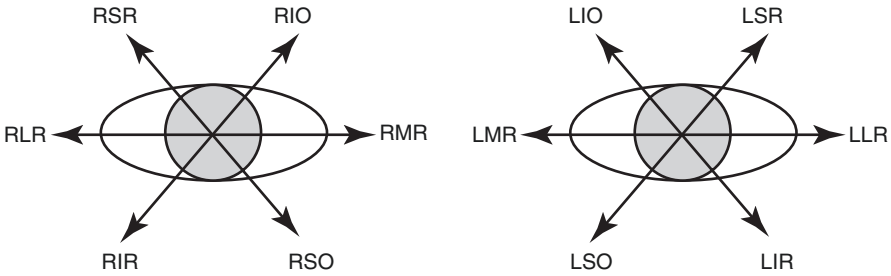


Fig. 4.1 Yoke muscles in six cardinal directions of gaze

one eye accompanies the other eye in all direction of gaze. Duane proposed the briefer term "comitant," which is now universally accepted in the literatures [1].

In a comitant strabismus, the amount of deviation remains the same in all direction of gaze. To maintain the comitance, the Hering's law of equal innervation, also called the law of motor correspondence, plays a major role [2]. According to this, the yoke muscles in each eye receive equal impulses to contract together to move the eyes toward the desired direction (Fig. 4.1). This comitance can be disturbed by many factors, which results in incomitant (noncomitant) strabismus. In an incomitant strabismus, the amount of deviation varies in different directions of gaze. Clinically, most incomitant strabismus are paralytic, restrictive, or of combined mechanism. Factors causing incomitance can be related to abnormalities of the extraocular muscles, orbit, neuromuscular junction, cranial nerves or their nuclei, supranuclear control, or combined mechanisms.

4.2.2 Primary Deviation and Secondary Deviation

A characteristic of incomitant strabismus is that the deviation varies with different fixating eye. The examiner measures the deviation, as is discussed in Chap. 2. It is to be remembered that the prism is not placed over the fixating eye. Using the measurement of exotropia as an example, when the examiner holds a base-in prism over the right eye, the right eye will be viewing in an abducted position as it looks through the prism. The left eye remains in its primary position during the measurement. Therefore, the left eye is the fixating eye during the measurement.

When the non-paretic or nonrestricted eye is used to fixate, the deviation measured is the primary deviation (measurement done with prisms over the primarily squinting eye). When the paretic or restricted eye is used to fixate (prisms over the normal eye), the deviation measured is the secondary deviation. In the condition of paralysis or restriction, the affected eye will need increased innervation to the paralyzed muscle to move to the fixating position. According to Hering's law, the increased amount of innervation will flow to the yoke muscle in the fellow eye, and this extra innervation will cause its excessive contraction exaggerating the strength disparity between these two yoke muscles. The result is an increased deviation when the affected eye is used for fixating. This is the reason why *the secondary deviation is always greater than the primary deviation*.

4.2.3 Spread of Comitance

The difference in primary and secondary deviation is most obvious when the paresis is recent. In long-standing paresis of one muscle, changes occur in other extraocular muscles, called "muscle sequelae." The development of muscle sequelae consists the following stages:

1. Overaction of its contralateral agonist (yoke muscle)
2. Overaction of the ipsilateral antagonist
3. Secondary underaction of the antagonist of yoke muscle

For example (Fig. 4.2), in a patient with a right lateral rectus paralysis, an esodeviation will occur which is largest in right gaze (dextroversion). According to Hering's law, the increased amount of innervation necessary to move the paretic right lateral rectus also flows to the left medial rectus (the contralateral agonist) and results in an overaction of left medial rectus. The right medial rectus (the ipsilateral antagonist) is acting unopposed and will be more or less overacting. However, according to Sherrington's law, the right medial rectus is actually receiving lesser inhibitional innervation than normal, and consequently lesser force is required to overcome the right lateral rectus (which is paralyzed) to fixate on a target. Then, according to Hering's law again, the innervation to left lateral rectus (the contralateral antagonist) will also be reduced. The left lateral rectus will be acting like a paralyzed muscle. This phenomenon is called "inhibitional palsy of the contralateral antagonist.". The esodeviation in left gaze (levoversion) would increase and match that in right gaze gradually. The difference between primary and secondary deviation would decrease, and the strabismus becomes concomitant, called "spread of comitance." This development of muscle sequelae occurs more often when the paralysis is long-standing and affects a strongly dominant fixating eye. This phenomenon often makes identification of the primary paretic eye difficult.

4.3 Clinical Approach to Incomitant Strabismus

It is crucial to differentiate paretic or restrictive strabismus from comitant forms of strabismus, not only because the principles of the management of incomitant strabismus are different from that of comitant strabismus, but also because an acquired paretic strabismus may be an initial sign of a sinister neurological abnormality.

When a paralytic strabismus is suspected clinically, the clinician needs to determine the possible underlying etiology. The causes may range from a rather benign microvascular event to a potentially dangerous intracranial lesion. The aim of clinical investigations is to identify the etiology and initiate timely intervention, if required.

The differentiation between acquired and congenital paralyses is essential. In recently acquired paralyses, patients often have symptoms of diplopia, or blurred vision, and are aware of change in head posture. While in congenital paralyses, patients usually adapt to their visual disorder, perhaps with development of suppression. Old photographs are helpful to clarify the time of onset of abnormal head

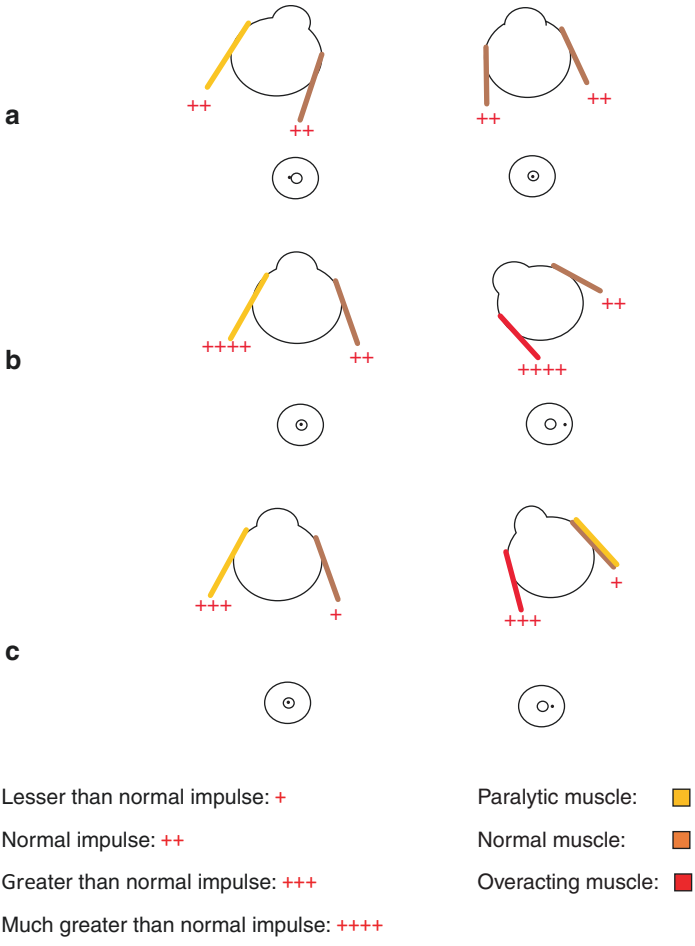


Fig. 4.2 Diagrammatic representation (in the presence of right lateral rectus (RLR) palsy) of the concepts of (a) *primary deviation* in the right eye (RE) which develops esotropia (ET) when normal impulse to contract (++) is reaching all muscles. (b) *Secondary deviation* in left eye (LE) when RE is forced to fixate with a much greater than normal impulse to contract (++++) reaching the paralyzed RLR and its yoke muscle, the left medial rectus (LMR). The normal LMR causes a much larger deviation in LE. (c) *Inhibitional palsy* of left lateral rectus (LLR) develops over time if the RE is dominant and prefers to fixate. The antagonist of the paralyzed muscle, i.e., RMR, starts receiving lesser impulse to contract as it is acting against a muscle (RLR) with reduced tonus. The same reduced impulse gets transmitted to the yoke of the antagonist of the paralyzed muscle, i.e., the LLR which starts underacting. Note that the secondary deviation reduces, compared to (b)

posture. Detailed history of antecedent events, including trauma, viral infection, meningitis, and microvascular risk factors such as diabetes mellitus, hypertension, and hyperlipidemia are important clues to potential causes.

The decision of whether to order neuroimaging in acquired paralyses is challenging to clinicians. In an era with many advances in treatment of stroke, neoplasm and

demyelinating diseases, early diagnosis of these conditions provides better clinical outcomes [3]. Multiple cranial nerve involvement and associated neurological symptoms and signs, such as headache, papilledema, and seizures, are indications for prompt neuroimaging. In isolated acquired ocular motor neuropathies, age of presentation and the cranial nerve involved are important factors for decision-making. Acute presentation in children almost always warrants a neuroimaging.

4.3.1 History Taking

The assessment starts with focused history taking as discussed in Chap. 2. The following information should be collected.

- The age of onset of the deviation. The circumstance under which the deviation becomes worse. Is it constant or intermittent? Is it unilateral or alternating? Is it worse at the end of the day?
- Associated symptoms, including blurred vision, double vision, eye strain, eye pain, watering, closing one eye in bright light, anomalous head posture, headache, dizziness, weakness and numbness of face, or extremities.
- Past ocular history, including glasses, patching, previous surgery, or trauma.
- Past history, including prematurity, developmental delay, hearing impairment, diabetes mellitus, hypertension, hyperlipidemia, trauma, or other systemic or neurological disorders.
- Family history, including ocular and systemic history, especially hereditary disorders.

Infants often have variable ocular deviation in the first month of life (oculomotor instability of infancy) [4]. The deviation of strabismus is nonstable until ~3 months of age. Infantile esotropia often occurs by 6 months of age. Accommodative esotropia typically occurs between 2 and 3 years of age. The age of onset for intermittent exotropia varies but is usually between 6 months and 4 years of age [5].

4.3.2 Head Posture

Anomalous head posture (AHP), compensatory head posture (CHP), or torticollis can result from ocular or non-ocular causes. Ocular AHP is often a compensatory response to minimize diplopia and improve the field of binocular vision in strabismus. The presence of AHP in a strabismus patient is suggestive of binocularity, and patching of one eye should eliminate it. In infantile nystagmus syndrome, AHP has been assumed to extend the foveation period and to adapt to the position of a strong dominant eye (Fig. 4.3) [6]. AHPs can be observed at any age, even as early as 6 months of age, when an infant just starts to sit unsupported. Prolonged AHP in children often results in facial asymmetry and influences musculoskeletal developments and should thus be corrected as early as possible.

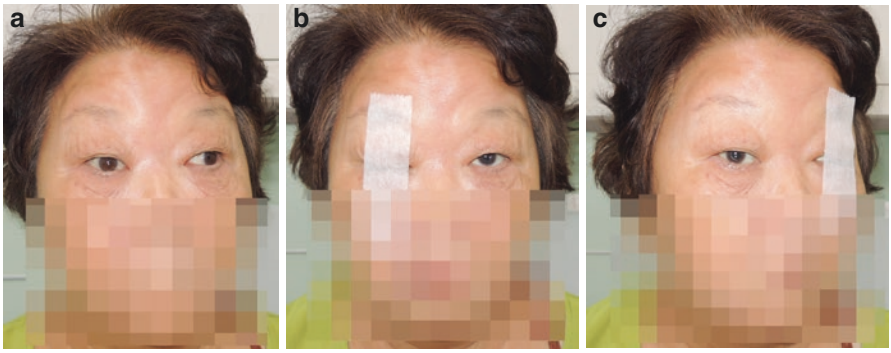


Fig. 4.3 Anomalous head posture (AHP) in a patient with congenital nystagmus with left exotropia (LXT) (**a**). The AHP is because of the fixating right eye (RE) occlusion of which eliminates the AHP (**b**); however, occlusion of the non-fixating left eye (LE) has no effect on it (**c**) (Note: This example should be considered as an exception rather than rule as in most patients with strabismus, AHP corrects with occlusion of either eye)

AHP has three components: (a) face turn, (b) head tilt, and (c) chin elevation or depression (Fig. 1.9). The axis of AHP is helpful in the diagnosis. A face turn may be associated with sixth nerve palsy (to the affected side), Brown syndrome (to the unaffected side), Duane syndrome, or homonymous hemianopia. A head tilt may be associated with superior oblique palsy (to the unaffected side), dissociated vertical deviation, or torsional nystagmus. A chin up/down may be associated with A and V patterns, thyroid eye disease, ptosis, Brown syndrome (chin up), or congenital cranial dysinnervation disorders. Patients with infantile nystagmus syndrome may have combined forms of AHP that include face turn, head tilt, and chin up/down to take advantage of the horizontal, torsional, and vertical components of their null points.

4.3.3 Ocular Movements

Ductions are monocular eye movements. Versions are conjugate binocular eye movements. To measure ductions, the examiner covers one of the patient's eyes and asks the patient to follow a fixation target with the other eye. To measure versions, the patients are examined with both eyes uncovered. In general, ductions are slightly greater than versions. If versions are normal, ductions are usually normal (see Chap. 1 also).

The examiner can follow a useful guideline to judge normal abduction and adduction. In full adduction, the nasal third of cornea is at the position of lower lacrimal punctum. In full abduction, the temporal corneal limbus touches the lateral canthus. Underactions are graded on a scale of 0 to −4, in which 0 indicates full ductions and −4 indicates no movement beyond the primary position. If the eye cannot even reach the primary position, a scale of −5 is recorded (Fig. 4.4).

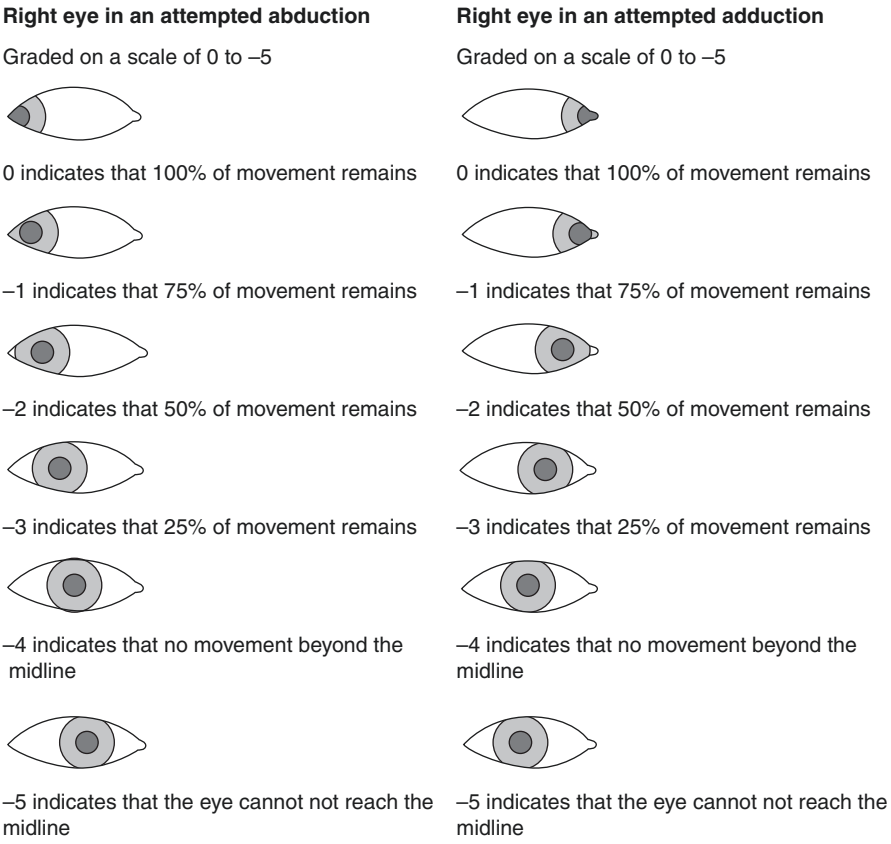


Fig. 4.4 Documentation of ocular ductions

Vertical ocular rotations are more complex as the vertical muscles have different secondary and tertiary actions as the globe rotates. However, an asymmetry of elevation (supraduction) straight upward or depression (infraduction) straight downward indicates possible muscle paresis or restriction. In full elevation, the lower limbus of cornea can pass above an imaginary horizontal line through lateral canthus. In full depression, the upper limbus of cornea can easily pass several millimeters below an imaginary horizontal line through lateral canthus, and the pupil gets covered by the lower lid. It is useful to record the limitation in ocular movements by photographs of the patient (Chap. 1).

When limitation of ductions is observed, it is necessary to perform *forced duction test* (FDT) to differentiate restriction from paresis. Office forced duction testing can usually be performed successfully in cooperative adults. The conjunctiva is anesthetized with topical 0.5% proparacaine eyedrops. An eyelid speculum is applied. The non-testing eye is covered. The patient is instructed to look as far as

possible in the direction of suspected limited duction. The examiner grasps the conjunctiva near limbus with fixation forceps and tries to rotate the globe further in the direction of limited duction. If the globe cannot be passively rotated further, restriction is present, and the result is recorded as positive. If further rotation is possible, the result is negative, and paresis may be the cause of the limited duction. FDT and force generation test (FGT) are discussed in Chap. 6.

The overaction of inferior oblique is often assessed during examination for versions. Overaction of the adducting eye is assessed, and the other eye (abducting eye) should be fixating during assessment. It should keep looking straight across the lateral canthus. The overaction is graded on a scale of +1 to +4, in which +4 indicates the corneal reflection is located at the lower limbus and +2 indicates the reflection located at midway between the pupil center and lower limbus.

4.3.4 Three-Step Test

In patients with vertical deviation in primary position, the three-step test is a helpful diagnostic algorithm to detect an isolated cyclovertical muscle palsy [7]. The results of this test may be invalid in multiple muscle paralyses, restrictive strabismus, dissociated vertical deviation, and underlying vestibular or supranuclear disease [8]. The test should not be attempted in patients with cervical spine instability.

The examiner determines (a) which eye is hypertrophic in primary position, (b) whether the deviation increases in dextroversion or levoversion, and (c) whether the deviation increases with right or left head tilt. It is useful if the examiner can identify the eye with primary deviation (eye with muscle paralysis) initially.

4.3.4.1 Step 1

The examiner measures the deviation at primary position and marks the suspected underacting muscle groups. For example (Fig. 4.5), if right hypertropia (or left hypotropia) is present at primary position, the suspected underacting muscle groups are inferior rectus and superior oblique of the right eye (RIR and RSO) and superior rectus and left inferior oblique of the left eye (LSR and LIO). If at this stage the paralytic eye can be confirmed by primary and secondary deviation, the suspicion shifts to two muscles only.

4.3.4.2 Step 2

The examiner then determines if the deviation increases in right or left gaze. If the vertical deviation is larger in left gaze, of the four suspected muscles, only two could be underacting in this position, i.e., RSO and LSR. Larger deviation occurs only when the weak muscle is forced to contract due to larger impulse being transmitted to its yoke muscle (Hering's Law). So the suspicion shifts to either RSO or LSR. Also, if the examiner had determined the eye with paralytic muscle earlier, the diagnosis is made at this stage itself.

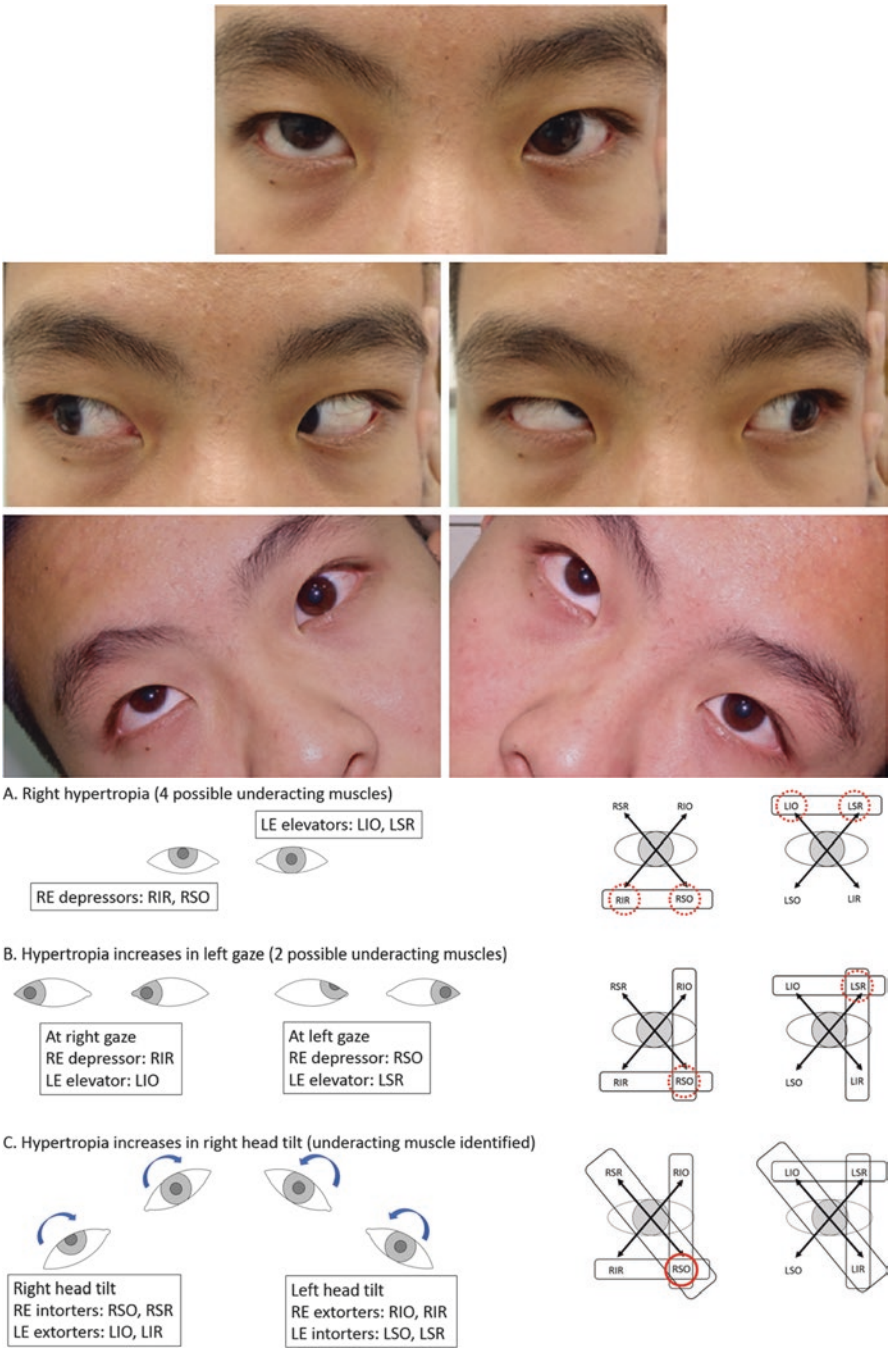


Fig. 4.5 Park's three-step test in right superior oblique palsy

4.3.4.3 Step 3

The examiner performs the Bielschowsky head tilt test by trying to estimate the change in deviation with the patient's head tilted to the right and left. Physiologically, a head tilt will induce compensatory ocular counter roll to reduce the torsions of the eyes relative to space. For instance, a right head tilt will induce incyclotorsion of the right eye and excyclotorsion of the left eye. The two muscles being considered are both intorters (Chap. 1), and right eye intorter (RSO here) would be forced to contract on right head tilt and the left eye intorter (LSR here) on left head tilt. So, if the deviation increases in right head tilt, the paralyzed muscle is RSO.

4.3.5 Hess Chart Test

Hess chart is a valuable tool for investigation of incomitant strabismus. By dissociating the two images of each eye, the test detects the position of fixating eye and non-fixating eye in different directions of gaze. The Hess chart test is a subjective test. It requires good vision in both eyes and foveal projection with normal retinal correspondence.

A standard Hess chart test consists a gray Hess screen with tangential patterns. The cardinal positions of eye movement are shown in red color, operated manually or electrically, sequentially. The patient wears a red and green goggle, with the red filter before the fixating eye. On the Hess screen, there is a central point, which is the primary position, surrounded by an inner field of eight points and outer field of 16 points. The test is performed at a distance of 50 cm. At this test distance, the points on the inner field represent the fixation at 15° from the primary position and the outer field representing the fixation at 30° from the primary position. For subtle underactions or overactions, which are hard to detect by the inner field, the outer field should be examined as the deviation will be more evident as the eye moves further to the field of action of the paralyzed muscle (Fig. 4.6).

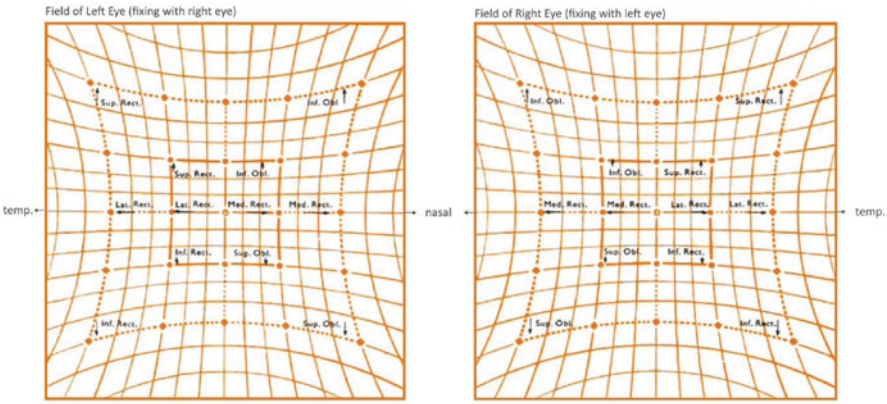


Fig. 4.6 Standard Hess chart

The results are plotted on Hess charts. The charts show deviation field for each eye when non-fixating. The field of left eye is recorded on the left chart and the field of right eye on the right chart. The recorded fields are direct recording of eye deviation. It means that a higher plot indicates a higher eye when the other eye is fixating. The Hess chart records the relative deviation of each eye, that is, a higher recording of one eye should be accompanied by a lower recording of the other eye, perhaps in different amplitude (due to difference in primary and secondary deviations), at the same fixation position.

The interpretation of Hess chart consists three important elements: position, size, and shape. The *position* of the field reflects the position of the eyes. A temporally displaced field reflects an exodeviation, and a nasally displaced field reflects an esodeviation. One should note that the plot of the central point is the deviation in the tested eye when the fixating eye is at the primary position and not the actual position of the tested eye.

The *size* of the field represents the amount of eye movement. An underaction of extraocular muscle will displace the plot interior to the normal field. Therefore, a reduced size indicates a limitation of movement. An overaction of extraocular muscle will displace the plot exterior to the normal field. Therefore, an enlarged size indicates an overacting movement. Comitance is indicated when the size of two field do not differ significantly.

The affected eye usually presents a smaller field. The examiner can identify the primary affected muscle, which shows the greatest underaction, in the eye with a smaller field (the affected eye) if the paresis is recently acquired (Fig. 4.7). The greatest overaction is demonstrated by the contralateral agonist (yoke) of the primary palsied muscle (Fig. 4.8).

Overaction is usually also shown by the ipsilateral antagonist of the primary palsied muscle usually. If the field is reduced at opposite directions, that is, underaction of a primary affected muscle as well as underaction of its ipsilateral antagonist, a mechanical restriction should be considered (Figs. 4.9 and 4.10).

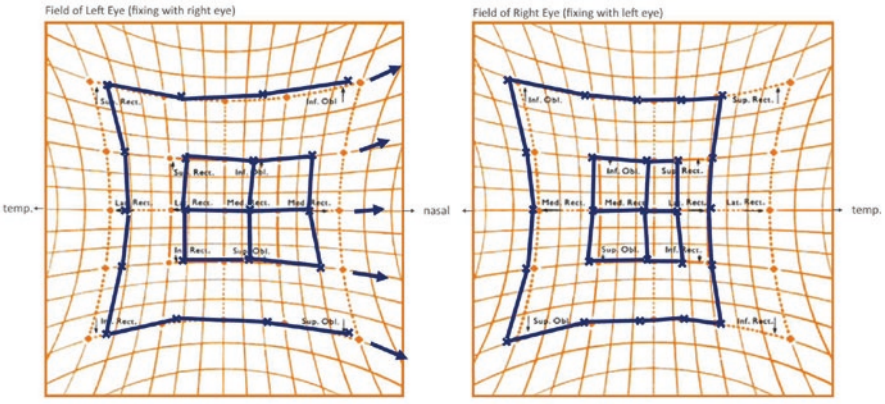


Fig. 4.7 Hess chart of right sixth nerve palsy

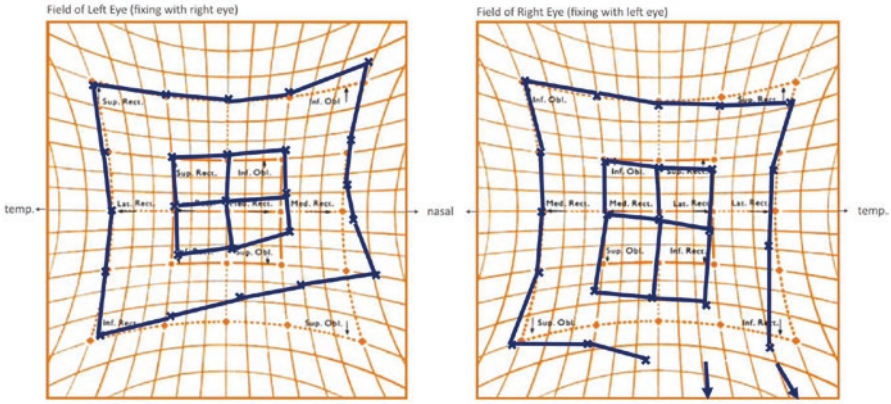


Fig. 4.8 Hess chart of left superior oblique palsy

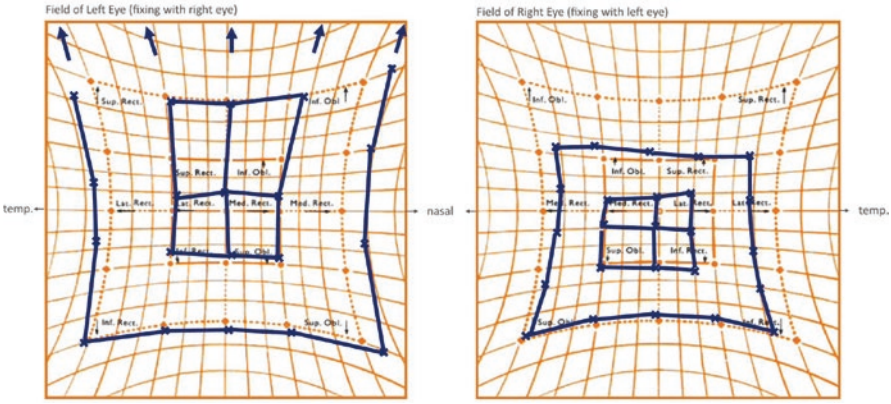


Fig. 4.9 Hess chart of thyroid eye disease with right inferior rectus fibrosis

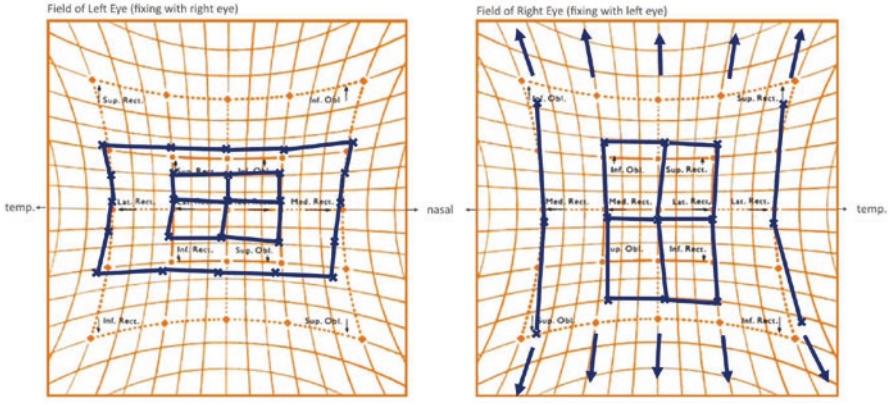


Fig. 4.10 Hess chart of left orbital floor fracture

The *shape* of the field implies the tilting of the field. It occurs when different amount of deviation is measured at different direction of gaze. A-V patterns are the most common examples of tilted shape of field. It should be noted that the tilted field does not indicate torsion.

4.3.6 Diplopia Charting

It is another simple yet effective test to evaluate incomitant strabismus. The test is performed using red and green glasses after eliminating any anomalous head posture of the patient. The details of this test are discussed in Chap. 2. Characteristic diplopia patterns also help in identification of the paralyzed muscle/s.

4.4 Salient Features of Individual Cranial Nerve Palsies

4.4.1 Sixth Nerve Palsy

Sixth nerve palsy is the most common cranial motor nerve palsy (Fig. 4.11). The patients commonly present with complains of sudden onset horizontal diplopia. Clinical findings include presence of face turn to the side of paralysis and esotropia with limitation of abduction in the affected eye. Uncrossed horizontal diplopia is characteristic in this condition. The anatomic position of the sixth cranial nerve makes it vulnerable to lesions from cerebellopontine angle and skull base. Simultaneous involvement of adjacent fifth, seventh, and eighth nerves is common. Neuroimaging is recommended for patients under 50 years of age, especially when multiple cranial nerves are involved. Isolated sixth nerve palsy in patients over 50 years of age with microvascular risk factors is often ischemic and

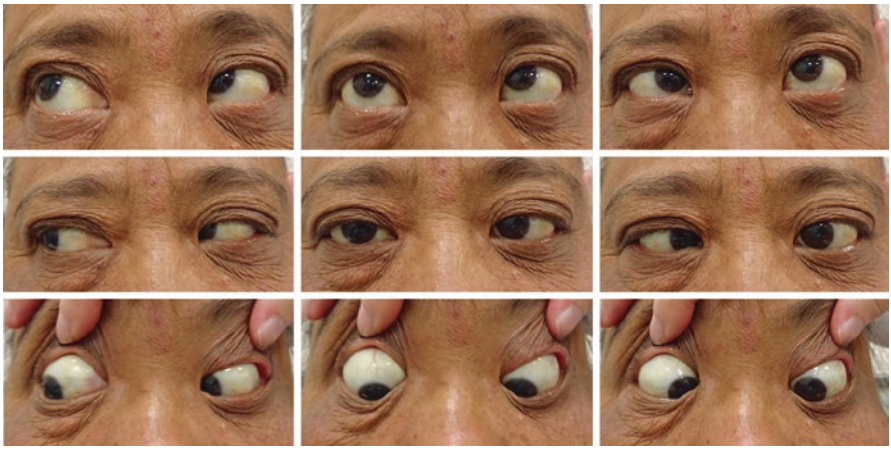


Fig. 4.11 Left sixth nerve palsy

can be followed for 3 months before neuroimaging [9]. Congenital sixth nerve palsy is rare in children. A congenital isolated abduction deficit in children should always raise suspicion of Duane syndrome. Neoplasms are the commonest cause of acquired sixth nerve palsy in children. Neuroimaging is thus recommended (Table 4.1).

4.4.2 Third Nerve Palsy

By virtue of extensive motor innervations of ocular structures, third cranial nerve palsy may have varied presentation. A complete third nerve palsy with pupil involvement typically presents with ptosis and dilated non-reacting pupil with eye in abducted and slightly hypotropic position (Fig. 4.12). A partial third nerve palsy may present with an isolated or multiple extraocular muscle palsy which may or may not involve the pupil. Among the extraocular muscles, isolated inferior oblique palsy is the rarest.

It is crucial to check the pupillary function when evaluating the third nerve palsy. The pupillomotor fibers of third nerve run on the dorsomedial surface of the nerve and are vulnerable to compression by aneurysm arising from posterior communicating artery (PcoA). A pupil involving third nerve palsy, complete or partial, is an indication for emergent neuroimaging to rule out the possibility of aneurysm.

When there is no evidence of pupillary dysfunction (pupil-sparing), the extent of third nerve palsy determines the next step. A pupil-sparing complete third nerve palsy in patients over 50 years of age most likely results from ischemic cranial neuropathy, especially when with the accompanying microvascular risk factors, such as diabetes mellitus, hypertension, or hyperlipidemia, exist. The palsy usually improves within 3–6 months. Neuroimaging is not required, except when the expected recovery does not occur or the palsy progresses or aberrant regeneration occurs in a nontraumatic palsy [21].

However, a pupil-sparing partial third nerve palsy has a different clinical meaning and often warrants imaging. Partial palsy may imply that some fibers of the third nerve are affected by a compressive lesion, and the other fibers, together with pupillomotor fibers, are not affected yet. The palsy may progress and later involve the pupil.

Another sign indicative of an underlying space occupying lesion is primary aberrant regeneration. Commonly, aberrant regeneration occurs after injuries when surviving nerve fibers grow to reinnervate the muscle and their misrouting occurs. However, when aberrant regeneration occurs in absence of trauma (in a presumed microvascular ischemic neuropathy), a compressive lesion should be suspected and neuroimaging should be obtained.

Third nerve palsy in children has different clinical features compared to adults. Commonest etiology is congenital, followed by trauma, viral infections, and tumor (Table 4.1). Aberrant regeneration is common in children. Pupil involvement is common in congenital form and does not suggest a compressive lesion as in adults [22]. Associated neurological anomalies such as arachnoid cyst and optic hypoplasia may exist. Neuroimaging is recommended in all children with third nerve palsy [7].

Table 4.1 Common causes of palsy of ocular motor nerves and indications for neuroimaging

	N III (Oculomotor)		N IV (Trochlear)		N VI (Abducens)	
	Children [7, 10–13]	Adults [14]	Children [15, 16]	Adults [17]	Children [18–20]	Adults [8]
Etiology (in decreasing frequency)	Congenital, trauma, infection, tumor	Aneurysm, microvasculopathy[a], trauma	Congenital, trauma, idiopathic	Congenital, trauma, microvasculopathy[a]	Tumor, hydrocephalus, trauma, infection	Microvasculopathy[a], trauma, tumor
Indications for neuroimaging	All patients	Pupil involvement, below 50 years (not at risk for microvasculopathy), non-resolving till 3 months, trauma, aberrant regeneration in non-traumatic palsy, partial palsy	Trauma	Acquired palsy below 50 years (not at risk for microvasculopathy), non-resolving till 3 months, trauma	All patients	Patients below 50 years (not at risk for microvasculopathy), non-resolving till 3 months, trauma

[a]Common microvasculopathies are diabetes mellitus and hypertension

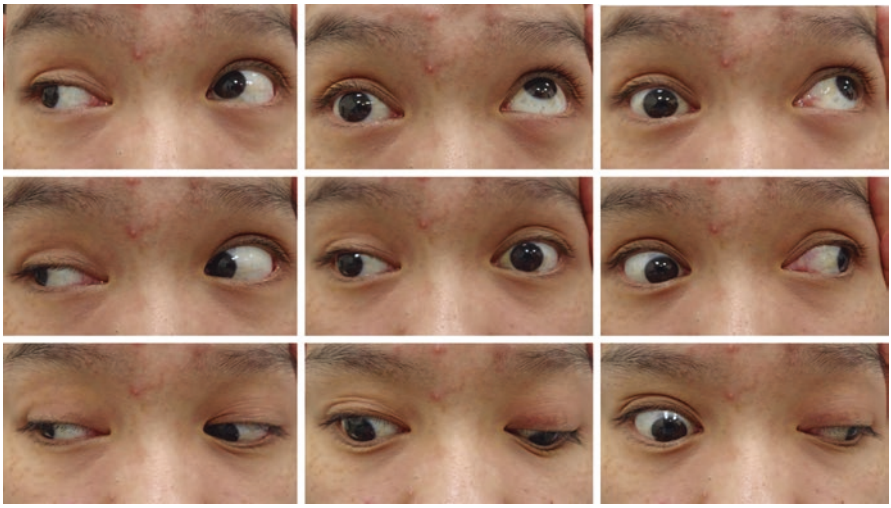


Fig. 4.12 Right third nerve palsy

4.4.3 Fourth Nerve Palsy

Fourth nerve palsies are usually congenital (Table 4.1, Fig. 4.13), and patients may be asymptomatic at early age. There can be associated anomalies of the nerve itself, tendon of superior oblique muscle, structure of trochlea, or insertion site on the globe.

Most of the acquired forms of fourth nerve palsy are caused by trauma or microvascular ischemia. Fourth nerve is the only cranial nerve exiting from dorsal midbrain and has the longest intracranial course. It is therefore vulnerable to closed head trauma. Patients with acquired palsy present with asthenopic symptoms, vertical diplopia, and AHP (head tilt to opposite side of palsy). Patients often complain of troublesome diplopia while climbing down the stairs.

Considering the low yield of neuroimaging in acquired non-traumatic fourth nerve palsies, patients over 50 years of age with microvascular risk factors can usually be followed for 3 months before neuroimaging [23].

4.5 Principles of Nonsurgical Management for Incomitant Strabismus

Nonsurgical management is aimed at preventing complications like amblyopia, contractures, etc. and making the patient comfortable till recovery occurs or definite surgical management is done. The patient is made comfortable by increasing the

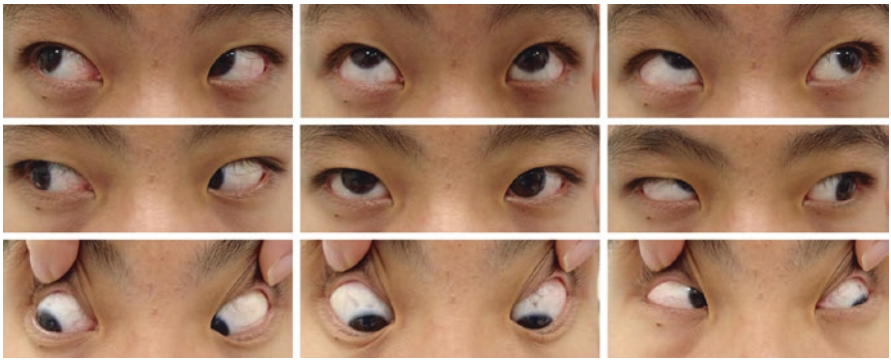


Fig. 4.13 Right fourth nerve paresis. Note the increased right hypertropia in levoversion and reduced motility of right eye in levodepression

Fig. 4.14 Segmental occlusion for diplopia in levoversion

diplopia-free field, eliminating head posture, and ensuring better cosmesis during the recovery. Nonsurgical options may be considered for long-term management in small-angle deviations specifically when the primary and downgazes are diplopia-free or when the surgery has to be delayed for various reasons. These are discussed in Chap. 5.

Children should be allowed to adopt a head posture as it allows binocular fusion. Alternate occlusion (daily closure of one eye alternately) prevents contractures and eliminates diplopia [17]. It also prevents amblyopia in small children. For small-angle deviations, Fresnel prisms may be tried; however, their utility is limited due to incomitance of deviations. Segmental occlusion of one eye in the direction of diplopia (Fig. 4.14) is also useful in selected cases [24].

In sixth nerve palsy, botulinum toxin is useful in normalization of head posture and increasing the diplopia-free field. It also prevents contractures, improves the patients subjectively, and is believed to reduce the length of morbidity [25–27]. Two to three units are injected in the antagonist or the yoke of the paralyzed muscle. Its utility is limited in third nerve palsy because of multiple extraocular muscle involvement. Botulinum toxin is discussed in Chap. 9.

4.6 Principles of Surgical Management for Incomitant Strabismus

To achieve a good clinical outcome, the surgeon must identify all underlying mechanisms of incomitant strabismus before surgical planning. The mechanisms may be restrictive, paralytic, or combined. Considerations about timing of surgery, procedure, and expected outcome differ for different disease entities. The family must be informed that expecting complete normalization of motility is unrealistic, and the surgery is targeted at restoring binocular single vision in primary and downgazes only.

4.6.1 Paralytic Strabismus

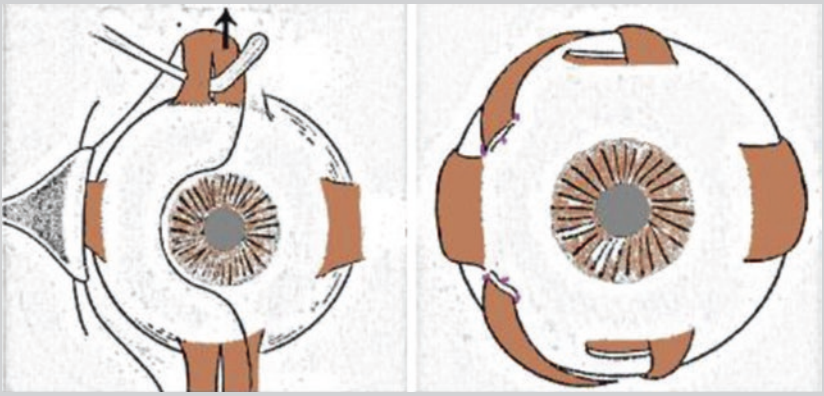
For acquired cranial nerve palsies resulting from trauma, compression or microvascular ischemia surgeries are usually withheld for a recovery period of 6 months after onset or even longer if the recovery is ongoing. In the recovery period, prisms, occlusion, or botulinum toxin are used as discussed in the previous section.

4.6.1.1 Sixth Nerve Palsy

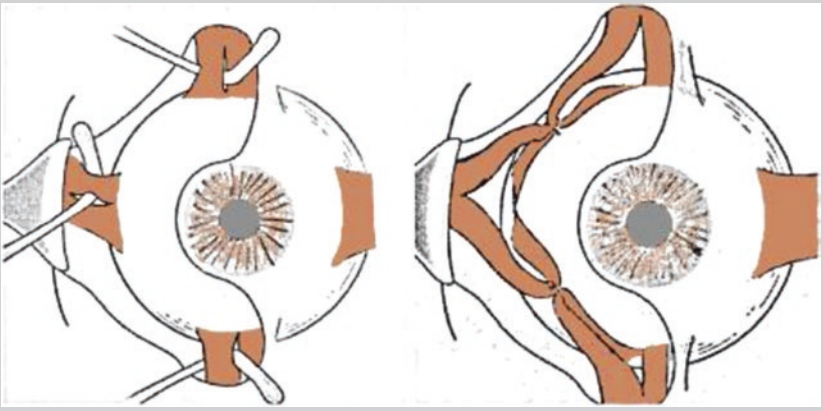
The surgical treatment of sixth nerve palsy depends on the amount of residual lateral rectus muscle function after recovery. Patients with good residual function have esotropia <20 PD in the primary position with almost full abduction, and those with moderate residual function have esotropia >20 PD with <−2 limitation of abduction. In these patients, ipsilateral medial rectus recession with lateral rectus resection is useful [14]. A posterior fixation suture (Faden) on the contralateral medial rectus (yoke) may be considered for deviation that is much larger in lateral gaze (direction of action of the paralyzed muscle). Patients with larger deviations over 40 PD in primary position may also require recession of the contralateral medial rectus as a staged procedure [28]. Patients with poor or absent residual function have −3 or greater limitation of abduction, some even unable to abduct past the midline. These patients have a negative FGT (discussed in Chap. 6). Resection of an almost fully paralyzed lateral rectus will have little effect, and vertical muscle transposition procedure, with recession of ipsilateral medial rectus, is indicated [29]. Considering the risk of anterior segment ischemia in surgeries involving multiple extraocular muscles, the split tendon transfer procedures are usually preferred than a full tendon transfer procedure [30]. See Text Box 4.1 [31–33]. These procedures are best performed by strabismologists.

Text Box 4.1: Selected Transposition Procedures

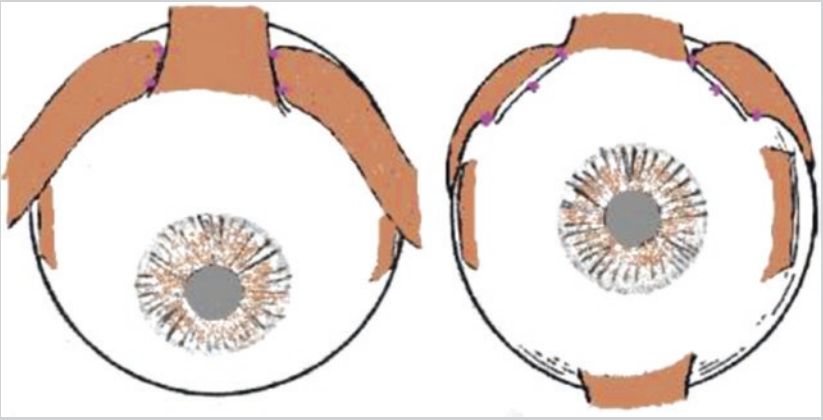
Partial tendon transposition (Hummelsheim) [11]: Temporal halves of SR and IR are transposed to LR insertion in LR palsy



Rectus muscle union (Jensen) [12]: In LR palsy, the SR, IR, and LR are split along their long axis. The lateral halves of SR and IR are united with superior and inferior halves of LR, respectively.



Full tendon transposition for vertical muscle palsy (Knapp) [13]: Both horizontal recti are transposed to the SR in double elevator palsy. The insertions may be placed parallel to the long axis of SR or following the spiral of Tillaux



4.6.1.2 Third Nerve Palsy

The surgical treatment of third nerve palsies depends on the number of muscles involved. For single rectus muscle involvement (medial rectus, superior rectus, inferior rectus), the same surgical principle discussed for sixth nerve palsy applies, that is, the amount of residual muscle function guides the decision to perform either recession-resection or muscle transposition. For multiple muscle involvement, surgical plan is decided after assessment of each muscle individually.

For complete third nerve palsies, there are only lateral rectus and superior oblique functioning. The coexisting paralyses of superior rectus, inferior rectus, and medial rectus make tendon transfers from adjacent muscles not feasible. Extreme large lateral rectus recession, sometimes with orbital wall fixation, has been proposed [34]. However, the lateral rectus often reattaches to the globe and creeps forward to result in recurrence of exotropia. Nasal transposition of split lateral rectus can be performed in cases without extensive scarring or lateral rectus contracture from prior muscle surgery [35].

The involvement of levator in third nerve palsy poses another important issue in treatment. The poor or absent levator function often requires frontalis sling procedure to correct the neurogenic ptosis. However, risk of exposure keratitis should be kept in mind in the absence of normal Bell's phenomenon because of paralyzed superior rectus. Some patients develop aberrant regeneration further complicating the situation.

Considering the possibility of troublesome postoperative diplopia and exposure keratitis, careful risk-benefit analysis should be done before initiating treatment [31].

4.6.1.3 Fourth Nerve Palsy

The surgical planning for fourth nerve palsy is complicated due to frequently late presentation with spread of comitance. A general rule is that the surgery is performed on the muscle with greatest overaction in the field of the largest vertical deviation on Hess Chart. Most patients of fourth nerve palsy manifest isolated ipsilateral inferior oblique overaction without obvious superior oblique underaction. Based on this principle, ipsilateral inferior oblique weakening is a frequently performed, effective, and safe initial procedure. Recession of contralateral inferior rectus or recession of ipsilateral superior rectus is performed for larger vertical deviations. When strengthening surgery of superior oblique is indicated, intraoperative exaggerated traction test is performed, and superior oblique tuck is done if tendon laxity is confirmed [36]. These procedures are again best performed by strabismologists.

4.6.2 Restrictive Strabismus

Mechanical restriction results from multiple factors often involving the muscle and its surrounding tissue. Thyroid ophthalmopathy and orbital trauma are the common causes. Surgical management of strabismus in thyroid ophthalmopathy is deferred until the disease is stable. In strabismus associated with trauma, early surgery is advised if significant soft tissue entrapment is visible on imaging.

When definitive surgery is undertaken after control of acute inflammation, preoperative evaluation with forced duction test is important to reveal limitation in duction. The restriction, if clinically significant, is relieved during surgery by release of the scarred or entrapped tissue or by muscle weakening procedure. However, it must be kept in mind that a muscle weakening procedure to correct alignment in the primary position may induce strabismus in the field of action of the weakened muscle which would subsequently require a posterior fixation suture on its yoke muscle [37].

4.6.2.1 Thyroid Eye Disease

Thyroid eye disease (TED) is a self-limited autoimmune inflammatory disorder. Most TED patients have a biphasic course, with an active or progressive phase for up to 18 months, followed by an inactive or stable phase. A characteristic fusiform swelling of the extraocular muscles belly with sparing of tendon on CT scan is often diagnostic. Severe exophthalmos, elevated intraocular pressure, and optic nerve compression may develop and require orbital decompression during the active phase. Strabismus surgery should be considered when the disease has stabilized for 4–6 months and should be delayed until after orbital surgery if necessary.

The surgery is aimed at restoring single binocular vision in primary and downgaze. TED often has both vertical and horizontal deviations (Fig. 4.15). Multiple surgeries are required and adjustable sutures should be used. Traditionally, only recession was recommended for TED; however, resections may also be done for large deviations [38]. Adjustable sutures are often necessary for severe restriction.

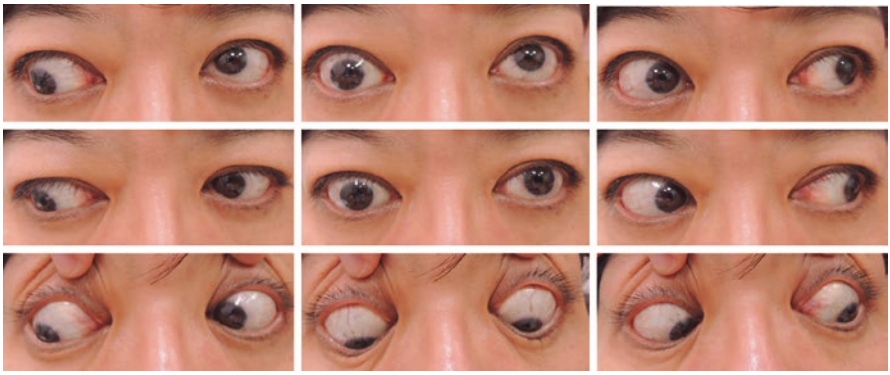


Fig. 4.15 Restrictive strabismus involving right inferior rectus in thyroid eye disease

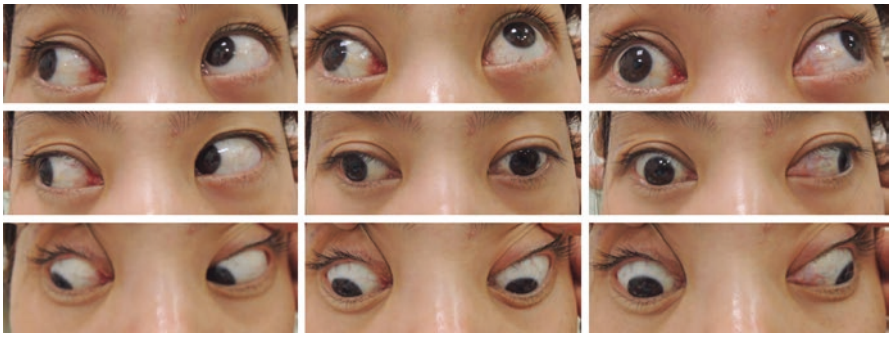


Fig. 4.16 Limitation of elevation and adduction of the right eye after fracture of the orbital floor

The aim should be slight under correction because of the late tendency toward overcorrection in TED [39].

4.6.2.2 Orbital Wall Fracture

The strabismus after orbital wall fracture can be caused by muscle injury, muscle entrapment, soft tissue edema, or hemorrhage (Fig. 4.16). Prompt computed tomography (CT) should be obtained to determine extent of orbital fracture and entrapment of ocular tissue. If CT shows clear evidence of extraocular muscle entrapment, early orbital surgery should be considered before permanent tissue changes. If there is no obvious entrapment, the patient may be managed nonsurgically (as discussed earlier). Should troublesome diplopia with no evidence of improvement persist at 6–8 weeks after trauma, strabismus surgery may be considered. However, it must be kept in mind that etiology of strabismus after orbital fracture may be restrictive, paralytic, or combined. The restriction may result from soft tissue fibrosis after original trauma or iatrogenically from orbital wall repair. Recession of the tight muscle and lysis of surrounding fibrotic tissue are usually required. For the paralytic component, the previously described principles for paralytic strabismus should be applied.

4.7 Summary

- It is essential to identify incomitant strabismus as it could be indicative of sinister conditions like intracranial space occupying lesion (SOL).
- Incomitant strabismus can be paralytic or restrictive. Often, both paralytic and restrictive mechanisms coexist in long-standing incomitant strabismus.
- Trauma, microvasculopathies (like diabetes mellitus), thyroid eye disease, viral infections, and SOLs are common causes of acquired incomitant strabismus.
- Larger secondary deviation, diplopia, and anomalous head posture are important clinical features of acquired incomitant strabismus.
- Neuroimaging is indicated in all children presenting with acute-onset strabismus and in adults with pupil involving complete and pupil-sparing *partial* third cranial nerve palsy. Neuroimaging should also be recommended in acquired strabismus in patients below 50 years, non-resolving till 3 months, or aberrant regeneration in non-traumatic palsy.
- Surgical correction is deferred till spontaneous recovery is occurring. Botulinum toxin injection and alternate occlusion may be prescribed during recovery. Surgery is planned to correct the deviation and enhance diplopia-free field in primary and downgazes.
- Choice of surgery is guided by residual motilit in the paralyzed muscle. Muscle transposition surgeries are indicated in completely paralyzed muscles. Adjustable sutures are often used to improve surgical outcomes.
- Early surgery may be needed when restrictive mechanisms are suspected as delay may lead to permanent alterations in extraocular muscle architecture.

4.8 Multiple Choice Questions

1. Which of the following condition is *not* an indication for emergent neuroimaging?
   (a) Pupil involving partial third nerve palsy in adults
   (b) Pupil-sparing complete third nerve palsy in adults
   (c) Acquired sixth nerve palsy in children
   (d) Acquired sixth and seventh nerve palsies in adults

Answer: (b) Pupil-sparing third nerve palsy in adults is usually due to microangiopathy occurring in diabetes mellitus or hypertension. All other conditions commonly occur in central nervous system neoplasms and warrant early imaging.

2. Which of the following is the procedure of choice for right superior oblique palsy with greatest deviation at left upper gaze?
   (a) Right superior oblique tuck
   (b) Right inferior oblique recession
   (c) Left inferior rectus recession
   (d) Right superior rectus recession

Answer: (b) The muscles maximally contracting in left upper gaze are right superior rectus and left inferior oblique (see Chap. 1). Right superior oblique palsy would cause hypertropia with inferior oblique overaction of the right eye. Inferior oblique recession would be indicated for its management.

3. A patient (Fig. 4.17) presented with right sided tilt of the head while walking. He complained of diplopia while going downstairs. All this happened after recovering from the head injury following road traffic accident. On examination there was hypertropia of left eye. The hypertropia worsened in right gaze, with improvement on right tilt. The probable diagnosis is:
   (a) Right SO palsy
   (b) Right SR palsy
   (c) Left SO palsy
   (d) Left SR palsy

Answer: (c) This is a case of acquired Left SO palsy. The test employed to clinch the diagnosis is "Park three-step test".

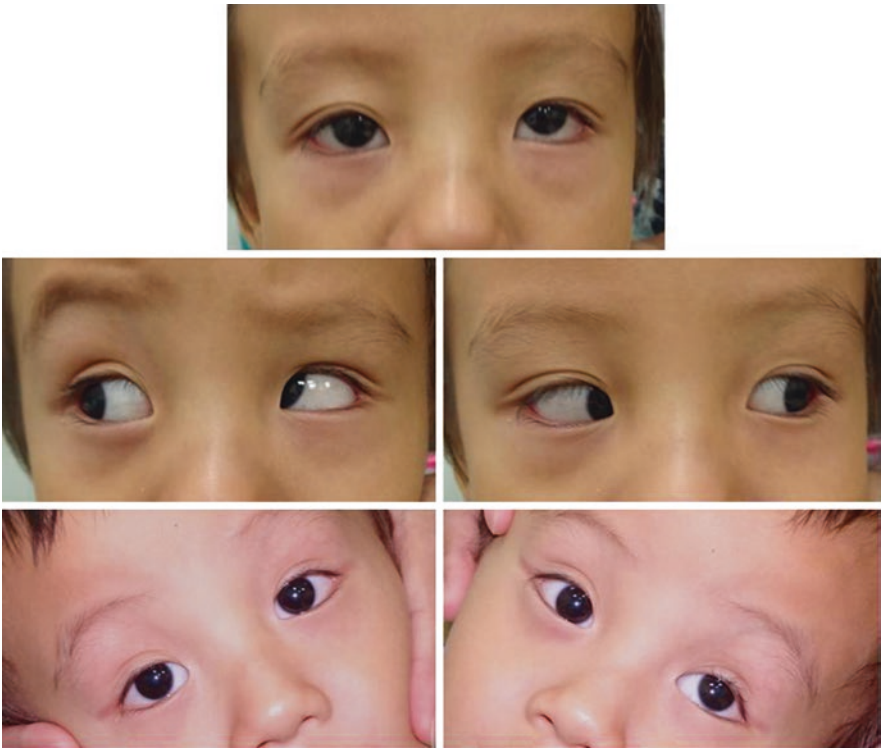


Fig. 4.17 Photography of the patient in Question 3

4. Late clinical findings consistent with an inferior blowout fracture of the orbit include all of the following *except*:
   (a) Proptosis
   (b) Ipsilateral hypotropia on upgaze
   (c) Ipsilateral hypertropia on downgaze
   (d) Positive forced ductions

Answer: (a) In the acute setting, there may be proptosis, but this usually gives way to enophthalmos as swelling subsides.

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