Understanding the Causes of Vertical Diplopia – Red Flag or Muscle Weakness

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(I) **Objectives:** Upon completion of this educational session, the participant will be able to:
- (A) Describe the neural pathway responsible for the ocular tilt reaction, including anatomy and biomechanics of the extraocular muscles, with related impairments associated with extraocular muscle weakness.
- (B) Measure ocular alignment using prism bars, double Maddox rod and phoria cards in unique head positions.
- (C) Discuss a new device that measures the perception of binocular alignment by providing discontinuous visual signals (the VAN and TAN Tests).
- (D) Know when to refer patients for formal ocular alignment testing and to modify oculomotor exercises due to ocular alignment.

(I) Titrated Approach to Diagnosing Pathologies Causing Dizziness and Vertigo Symptoms[3]
- (A) Timing
  - (1) Episodic vestibular symptoms
  - (2) Acute vestibular symptoms
  - (3) Chronic vestibular symptoms
- (B) Triggers
  - (1) Physical – precipitated by some specific obligate action or event such as head motion, change in body position, loud sounds, Valsalva maneuvers, etc.
  - (2) Spontaneous - unprovoked
- (C) Targeted History/Examination
  - (1) Dix-Hallpike Test
  - (2) HINTS-plus.[4] HINTS without or with hearing loss.
    - a. Head Impulse Test
    - b. Directional Changing Nystagmus
    - c. Skew Deviation – 100% sensitive and 96% specificity.
- (D) Acute Vestibular Symptoms. Differentiate central v peripheral vestibular lesion. Findings suggest central lesion if (1) INFARCT – impulse normal, fast phase alternating, refixation on cover test or (2) HINTS Plus – HINTS with hearing loss.[3]

(II) Anatomy and Physiology of Acquired Binocular Misalignment.
- (A) Oculomotor system is divided by anatomical location.
  - (1) Nuclear – Cranial Nerve nuclei (III, IV, VI, VIII)
  - (2) Infranuclear – fascicles and nerve fibers (CN III, IV, and VI) and extraocular muscles.

(3) Internuclear – Medial longitudinal fasciculus (MLF). Connections between oculomotor, trochear, abducens and vestibular nuclei.
(4) Supranuclear – cortical structures and pathways that descend into the brainstem and are rostral to the ocular motor nuclei.

(B) Infranuclear and nuclear control of eye movements – extraocular muscles and innervation. Eye movements are produced by 6 extraocular muscles. The muscles work together in 3 pairs to move the eye in the horizontal, vertical, oblique and rotary planes.

(1) Medial rectus (MR) and lateral rectus (LR) adduct and abduct the eye in all positions of horizontal gaze.
(2) Superior Rectus (SR)(Figure 1)[1]. Courses forward at an angle of 23° to the medial wall of the orbit and attaches to the globe superiorly.
  a. In the primary position, the movement is combined elevation and intorsion plus slight adduction (1B).
  b. With the eye abducted 23°, the plane of the SR parallels the vertical anterior-posterior plane of the globe. The movement is primarily elevation (1C).
  c. With the eye adducted, the plane of the SR becomes perpendicular to the visual axis. The movement is primarily intorsion (1A).

(3) Superior Oblique Muscle (SO) (Figure 2)[1]. The tendon reflects back from the superior-medial wall of the orbit at an angle 51°with the medial wall of the orbit and inserts into the posterotemporal sclera.
  a. In the primary position, the movement is combined intorsion and slight abduction (2B).
  b. With the eye adducted, the plane of the SO tendon parallels the visual axis and pulls the eye downward (2A).
  c. In 39° of abduction, the visual axis is perpendicular to the SO plane and the movement is intorsion (2C).

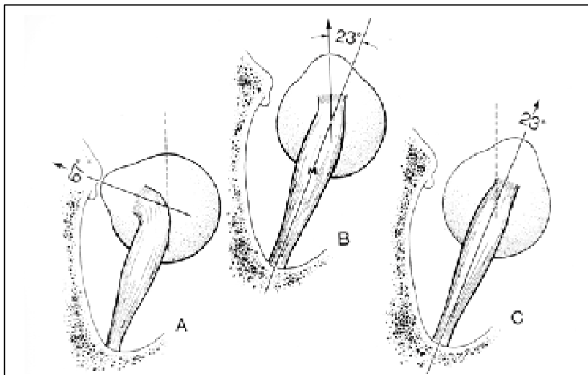

Figure 1. Anatomy of Superior Rectus Muscle[1]. Different starting positions: A. Adducted. B. Primary position. C. Abducted 23°.

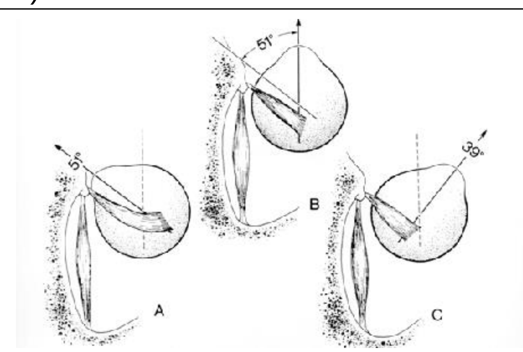

Figure 2. Anatomy of Superior Oblique Muscle[1]. Different starting positions: A. Adducted. B. Primary position. C. Abducted 39°.

(4) Inferior rectus (IR). Angle of pull of tendon on eye similar to superior rectus but reversed, Table 1

(5) Inferior oblique (IO). Angle of pull of tendon on eye similar to superior oblique but reversed, Table 1.

Table I. Innervations and Actions of Extraocular Muscles				
Nerve	**Muscle**	**Primary Position**	**Adduction**	**Abduction**
III	Superior Rectus	**Elevation**, Incyclotorsion, adduction	**Intorsion**, adduction, elevation	**Elevation**
III	Inferior Oblique	**Extorsion**, elevation, abduction	**Elevation**, abduction, extorsion	**Extorsion**, abduction, elevation
III	Inferior Rectus	**Depression**, extorsion, adduction	**Extorsion**, adduction, depression	**Depression**
IV	Superior Oblique	**Intorsion**, Depression, abduction	**Depression**, abduction, intorsion	**Intorsion**, abduction, depression
VI	Lateral Rectus	Abduction	Abduction	Abduction
III	Medial Rectus	Adduction	Adduction	Adduction

(C) Nuclear and infranuclear. Final common pathway – innervation to eye muscles.
- (1) Oculomotor Nucleus (CN III). Compartmentalized and organized by muscle action – SR, IR, IO, and MR.
- (2) Trochlear nucleus - SO (CN IV).
- (3) Abducens Nucleus - LR (CN VI).

(D) Organization of eye movements.
- (1) Midbrain center for control of vertical eye movements and vergence (also superior temporal sulcus).
- (2) Pons center for control of horizontal eye movements.
- (3) Lesion. Deficits ipsilateral to side of lesion.

(E) Insertions of extraocular muscles aligned with semicircular canal planes.
- (1) Lateral canal – horizontal eye movement.
- (2) Anterior canal – upward eye movement.
- (3) Posterior canal – downward eye movement.

(F) Insertions of extraocular muscles associated with Otolith stimulation– torsional eye movement.

(G) Function of the Vestibulo-Ocular Reflex. Hold image stable on the retina during brief head movements by generating a compensatory eye movement at the same velocity as the head but in the opposite direction. Two types of head movements – rotation and translation.

(H) **Canal receptors** contribute to 2 major functions:
- (1) Angular VOR (aVOR) - Rotation detected by canals and quantified by:
  - (a) VOR gain = eye velocity/head velocity or output/input
  - (b) Phase = temporal difference between head and eye movements.
- (2) Vestibulocollic reflex – head righting. Mediated by medial vestibulospinal tract (Figure 3).
  - (a) First order neuron – receptors cristae ampullaris of canal.
  - (b) Second order neuron – Medial vestibular nucleus. **Descends bilaterally** to alpha motor neuron within spinal cord.
  - (c) Third order neuron – alpha motor neuron spinal cord.

(I) **Otolith receptors** contribute to 4 major functions:
  (1) perception of spatial orientation (earth verticality)
  (2) generation of the translational VOR (tVOR) during lateral, vertical, and fore-aft motion of the head
  (3) generation of ocular counter roll during static head tilt (torsional VOR).

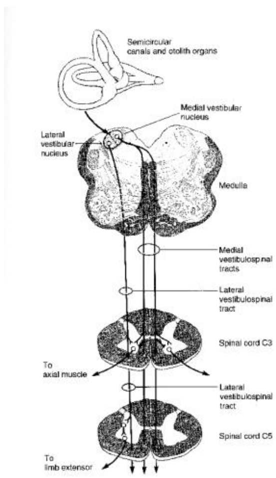

Figure 3. Lateral and Medial Vestibulospinal Projections.

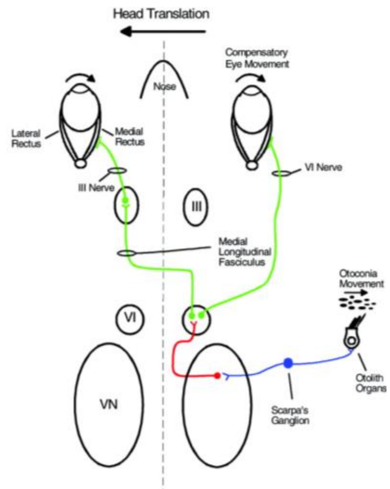

Figure 4. Translational VOR[5].

  (4) Generation of protective extension – lateral vestibulospinal tract. Second order neuron **descends ipsilaterally** to alpha motor neuron within spinal cord.

(J) **Physiologic Head Tilt**. Normal response to head tilt – generation of torsional VOR. With respect to the head tilt, the ipsilateral eye will elevate and counter roll (left) while the contralateral eye will depress and counter roll (left) in order to counter the change in head position and maintain binocular vision. Example – head tilt right – ocular counter roll towards the left.
  (1) Pathway (Figure 4).
    (a) First order neuron (blue) – receptors otolith organ.
    (b) Second order neuron (red) – vestibular nuclear complex.
    (c) Third order neuron – (green) CN III, IV, VI

(II) Superior Oblique Palsy (SO) results in symptom of vertical diplopia (Table I).
  (A) Signs (presumes unopposed activity of the palsied SO muscle's antagonist the inferior oblique):
    (1) central gaze ipsilesional hypertropia and extorsion of eye.
      (a) Greater in contralesional then ipsilesional gaze.
      (b) Greater in ipsilesional then contralesional head tilt.
    (2) Monocular tilt of subjective visual vertical.
  (B) Compensation.
    (1) Head tilt away from side of lesion and chin tuck to compensatory for vertical diplopia.

(III) Altered Gravity and the Effects on Binocular Misalignments. Unilateral lesion to the graviceptive vestibular pathways in the roll plane results in vestibular tone imbalance that causes perceptual tilt, head and body tilt, vertical misalignment of the visual axes (skew deviation) and ocular torsion.

(A) **Tilt of perceived vertical – subjective visual vertical (SVV)**. The patient's visual world is tilted with respect to the true earth vertical. For example, a reduced firing rate of the right utricle results in the brain perceiving the head is tilted to the left (error signal). The head reflexively tilts towards the right resulting in a compensatory head righting response[7].

(1) Graviceptive pathway[6]:

(a) Peripheral/lower brainstem: Lesion utricle and vertical canal ipsiversive SVV.

(b) Lower brainstem: Lesion vestibular nuclei (superior and medial) ipsiversive SVV.

(c) Midbrain: Lesion medial longitudinal fasciculus (MLF) in pontine and pontomesencephalic brainstem and Interstitial nucleus of Cajal (INC) induces contraversive SVV.

(d) Thalamus and cortex: variable, dependent on lesion above or below decussation.

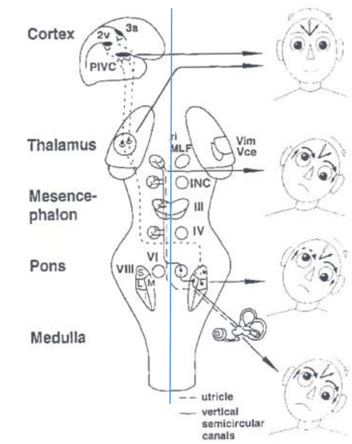

Figure 5. The effects of a lesion to the graviceptive vestibular pathways [6]. Pathway crosses at the pontine level. Ponto-medullary lesions below the crossing tilt effects ipsiversive and ponto-mesencephalic lesions above the crossing tilt effects contraversive.

(2) ***The perception of vertical provides the driving force*** for all components of the ocular tilt reaction - to realign the eyes and body with the perceived vertical[7].

(3) SVV tilts are the most sensitive sign of a vestibular tone imbalance in roll[8].

(4) ***SVV tilts found with both binocular and monocular viewing[9]***.

(B) **Compensation.** ***Ocular Tilt Reaction (OTR) –*** Pathophysiologic righting response – trying to realign the vertical axes of both the head and the eyes to the internal estimate of absolute earth -vertical[10](Figure 5 and Table 2). Lesion may result in complete or in single components of the OTR – skew deviation, ocular torsion, and head tilt. Pathway crosses at the pontine level. Ponto-medullary lesions below the crossing tilt effects ipsiversive and ponto-mesencephalic lesions above the crossing tilt effects contraversive. The ocular tilt reaction and its associated skew deviation tend to recover spontaneously over a median time of 7.5 months[11].

(1) ***Head tilt –*** otolithic tone imbalance medial vestibulospinal tract. The pathologic head tilt is ipsilateral to the hypotropic eye.

(2) ***Skew deviation –*** vertical misalignment of visual axes that does not map to cyclovertical eye muscles – one eye hypotropic. Caused by lesion in the brainstem, cerebellum, or peripheral vestibular system. Skew deviation is always combined with ocular torsion[12].

(a) May be comitant (the size of the hyperdeviation remains about the same in each field of gaze) or incomitant (skew deviation may be greater in one horizontal field of gaze and minimal in the opposite field of gaze). Usually comitant results in a negative Three Step Test (Park's test or Bielschowksy's head tilt test)[7].

(3) ***Ocular torsion –*** lack of ocular counter roll (OCR) or torsional VOR. The tilt orientation is always in the same direction as the observed ocular torsion[7].

(a) ***Normal ocular counter roll*** – upper poles of both eyes rotate in the opposite direction to the static head tilt. OCR compensates for 10-20% of the head tilts.

Tilt of head towards the right activates the right superior oblique and superior rectus muscles causing the right eye to incyclotort and elevate slightly and activates the left inferior oblique and inferior rectus muscles causing the left eye to excylotort and depress slightly.

(b) ***Pathological ocular counter roll*** – the upper poles of both eyes rotate in the same direction as the head tilt (hypotropic eye is excyclotorted and hypertropic eye incyclotorted).

(c) Ocular torsion may be conjugate or dissociated between the 2 eyes or present in one eye only[10].

(d) Marked decrease in static OCR gains with a mean reduction of 45% across both eyes and both directions[13]. OCR gains were asymmetrical between eyes and between torsional directions in about 90% of patient with brainstem or cerebellar lesions.

Table 2. Summary - Comparison Ocular Tilt Reaction v Superior Oblique Palsy

Ocular Tilt Reaction	Superior Oblique Palsy
Intorsion of higher eye/extorsion of the lower eye	Extorsion of higher eye
Binocular tilt of subjective visual vertical	Monocular tilt of subjective visual vertical
Head tilt compensatory for altered subjective visual vertical	Head tilt compensatory for vertical diplopia

(IV) The Process of Differential Diagnosis – Measurement of Ocular Alignment in the Clinic.

(A) History taking.
  (1) Is your double vision present in straight-ahead gaze?
  (2) Do the images separate more when you look to your right or left?
  (3) If it worsens in right gaze, does it worsen in gaze right and up or gaze right and down?
  (4) What happens if you tilt your head to the right or left shoulder?

(B) Observation
  (1) Abnormal head postures. The patient may adopt a compensatory head posture to optimize binocular single vision.
    a. Historical photo such as Driver's License
    b. Silhouette Posture grid
  (2) Lid abnormalities
  (3) Position of eye
  (4) Size of Pupil – pupillary light reflex. Swinging light test (Figure 6). Light shone through one pupil causes both sphincters to contract equally. The response to the illuminated eye is the direct reflex and the equal response of the un-illuminated eye is the consensual reflex. Reaction of pupil to light and video-oculography.
    a. Signs of optic nerve or afferent damage.
      i. Equal pupils.
      ii. No direct response.
      iii. Intact indirect response.

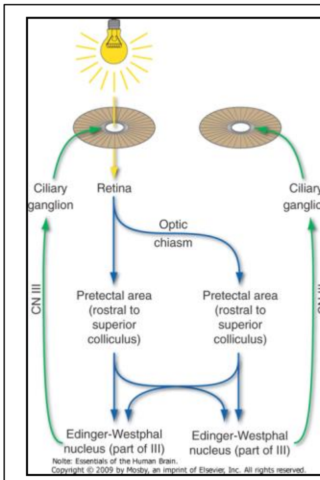

Figure 6. Circuitry of the pupillary light reflex.

b. Signs of oculomotor nerve or efferent damage.
   i. Dilated ipsilateral pupil.
   ii. Does not response to light shone into either eye (No direct or indirect response).

(C) Measurement of subjective visual vertical or tilt of perceived vertical. Vestibular imbalance in roll plane. Tests otolithic function and central graviceptive vestibular pathways in the roll plane. SVV tilt observed in 94% of patients with acute unilateral brainstem lesions[8] and > 80% of acute vestibular neuritis.[14]

(1) Bucket Test.[14] How to make your own bucket. http://www.mvrc.pitt.edu/files/SVV-Bucket_How-to.pdf.

(2) Monocular and binocular measurement of subjective visual vertical (Table 3).
   a. Healthy subjects align the bar within 1-2.5° from true vertical.[14]
   b. Subjects with central and peripheral pathology align the bar > 2.5°from true vertical.[14]

Table 3. Monocular and binocular measures of subjective visual vertical.[14]

Bucket Tests	SVV Monocular Left	SVV Monocular Right	SVV BInocular
Normal n = 30	1.2±.7	1.0±.8	.9±.7
Acute peripheral or central vestibular disorder n = 30	8.9±5.2	8.4±4.8	8.3±5

(D) **Measurement of Phoria/Tropia**. A complete assessment for binocular vision includes a minimum of 4 tests: near point convergence, cover test at distance and near, step vergence ranges at distance and near, and stereopsis testing.[15] We are discussing one of the four tests, cover test used to measure alignment of visual axes.

(1) Definitions:
   a. Strabismus – misalignment of the visual axes.
      i. Tropia. Misalignment of the visual axes during binocular viewing of a single target. Manifest deviation – present all the time. Focusing on target can't correct.
      ii. Phoria. Misalignment of the visual axes during monocular viewing of a single target. Latent deviation – deviation not always apparent. Observed with fatigue and when fusion (binocular vision) is broken.
   b. Deviation of visual axes. Tropia and Phoria may be:
      i. Exo – turned out.
      ii. Eso – turned in.
      iii. Hypo – turned down.
      iv. Hyper – turned up.
      v. Excyclo – torsional turn, upper pole templeward. Difficult to observe without ophthalmoscope.
      vi. Incycle – torsional turn, upper pole nasalward. Difficult to observe without ophthalmoscope.

(2) **Cover-Uncover Test** – near, intermediate, and far. Used to determine if ocular deviation is heterotropia or heterophoria. Provides information about the size and direction of the deviation in addition to the stability, constancy, laterality or control of a deviation. Disadvantage – can't detect very small deviations (up to 2-3 diopters).[16]

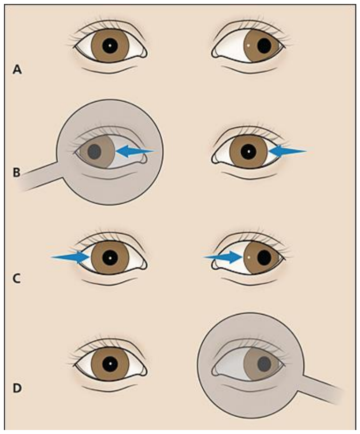

Figure 7. Cover-Uncover test. Exotropia.

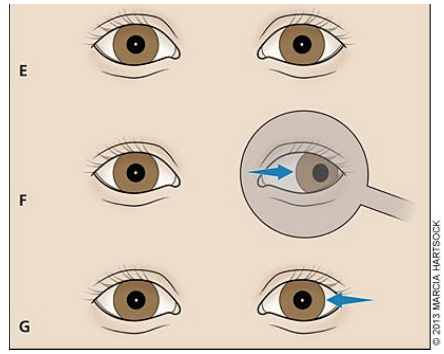

Figure 8. Cover-Uncover test. Exophoria.

a. Prerequisites for a reliable Cover Test.
    i. Eye movement range
    ii. Image formation and perception
    iii. Foveal fixation in each eye
    iv. Eliminating accommodative influences
    v. Attention and cooperation.
b. Test Procedures. Keep the room lights on. Tell patient you want to "find out how well your 2 eyes work together.
    i. Observe ***fixation stability*** for 5-10 s. Instruct patient to look at targets.
        1. Near cover test – eyes should be slightly downwards, fixation stick with letters of pictures 25-40 cm away. Target should be easy to read. Requires accommodation.
        2. Far target – head erect, eyes primary position of gaze, target 6 meters away. Target should be easy to read with both eyes. Requires accurate fixation.
    ii. Perform the cover/uncover test to look for a ***heterotropia*** – observe response of right eye and then left eye. Identifies tropia of uncovered eye (Figure &). Eye should be occluded at least 2 s.
        1. Response of right eye –
            a. Cover left eye, look for response of right eye.
            b. Cover right eye, look for response of left eye.
        2. Response of left eye –
            a. Cover right eye, look for response of left eye.
            b. Cover left eye, look for response of right eye.
    iii. If no heterotropia, look for ***heterophoria*** – observe the response of the covered eye as the cover is removed (movement of redress). (Figure 8)
        1. Deviations may be measured by placing prisms of increasing power in front of one eye until no movement is observed.

iv. If no heterotropia, perform the alternating cover test. Place the occluder before 1 eye for 2-3 s and then transfer quickly the occlude to the other eye, without pausing. Deviation will be seen as a re-fixation eye movement, when the cover is transferred from one eye to the other.
   1. Deviations may be measured by placing prisms of increasing power in front of one eye until no movement is observed.
   2. Observe the latency and the speed of the fusional recovery movement on uncovering – gives clues to strength of fusion reflex.

v. If no heterophoric movements are seen during the alternating cover test, perform the ***subjective cover test***.

(3) Maddox Rod Testing. Near does not take into account accommodation.

a. Prism. Neutralization of deviation with prisms by optically moving the image onto the fovea.
   i. Triangular or wedge shaped piece of refracting material. The base is thickest edge of the prism and the apex is the thinnest edge of the prism.
   ii. A prism changes the direction of light without changing its focus.
   iii. Measured in units called prism diopters (pd). 1 pd equals a 1 cm deviation at 1 meter.
   iv. In viewing an object through a prism, the image is displaced towards the apex.
   v. On the retina, the image is moved towards the base.

b. Maddox Rod Test Procedures. Dim the room lights.
   i. Horizontal heterophoria (horizontal alignment).
      1. Place the Maddox rod in front of the right eye (convention) making sure that the “grooves” are horizontal. The patient will see a vertical line through the ***right*** eye.
      2. Shine the light from a penlight. Penlight positioned 18 cm away from the bridge of the nose. The light will be seen with the ***left*** eye.
      3. Ask where light falls in relation to red line (Figure 9, 10, 11).
      4. Neutralization of deviation. Position horizontal prism bar in front of left eye in frontal plane position to neutralize deviation. Note diopters required to neutralize the deviation.

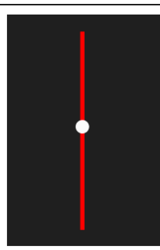

Figure 9. Orthophoria. Red line is seen straight through the light.

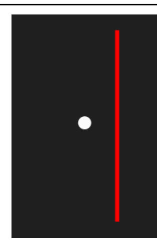

Figure 10. Esodeviation. Homonymous displacement or uncrossed diplopia. Red line is seen to the right of the light.

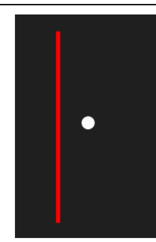

Figure 11. Exodeviation. Heteronymous displacement or crossed diplopia. Red line is seen to the left of the light.

   ii. Vertical heterophoria (vertical alignment).

1. Place the Maddox rod in front of the right eye (convention) making sure that the “grooves” are vertical. The patient will see a horizontal line through the ***right*** eye.
2. Shine the light from a penlight. Penlight positioned 18 cm away from the bridge of the nose. The light will be seen with the ***left*** eye.
3. Ask where light falls in relation to red line (Figure 12, 13, 14).
4. Neutralization of deviation. Position vertical prism bar in front of left eye in frontal plane position to neutralize deviation. Note diopters required to neutralize the deviation.

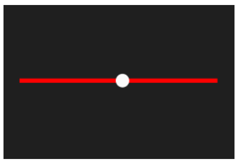

Figure 12. Orthophoria. Red line is seen straight through the light.

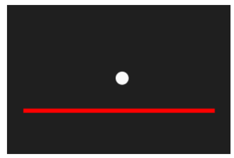

Figure 13. Hyperdeviation. Red line is seen below the light.

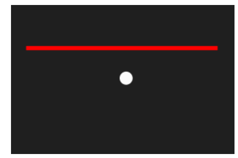

Figure 14. Hypodeviation. Red line is seen above the light.

iii. Consider referral.

1. Horizontal balance - Esophoria 10 diopters or more and Exophoria 8 diopters or more
2. Vertical balance - 2 diopters or more

(4) **Phoria cards near and far**. Near takes into account accommodation.

a. Maddox Rod Test Procedures. Dim the room lights.

i. Horizontal heterophoria (horizontal alignment).

1. Place the Maddox rod in front of the right eye (convention) making sure that the “grooves” are horizontal. The patient will see a vertical line through the ***right*** eye.
2. Shine the penlight through the central aperture of the phoria near/distant card. Penlight positioned 16”/10’ away from the bridge of the nose. The light will be seen with the ***left*** eye.
3. Ask where light falls in relation to red line (Figure 15, 16, 17).
4. Neutralization of deviation. Position horizontal prism bar in front of left eye in frontal plane position to neutralize deviation. Note diopters required to neutralize the deviation.

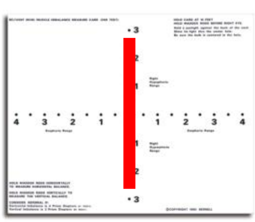

Figure 15. Orthophoria. Red line is seen straight through the light.

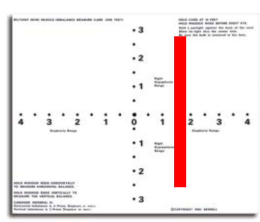

Figure 16. Esodeviation. Homonymous displacement or uncrossed diplopia. Red line is seen to the right of the light.

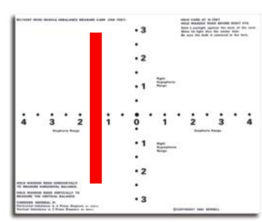

Figure 17. Exodeviation. Heteronymous displacement or crossed diplopia. Red line is seen to the left of the light.

ii. Vertical heterophoria (vertical alignment).

1. Place the Maddox rod in front of the right eye (convention) making sure that the "grooves" are vertical. The patient will see a horizontal line through the ***right*** eye.
2. Shine the penlight through the central aperture of the phoria near/distant card. Penlight positioned 16"/10' away from the bridge of the nose. The light will be seen with the ***left*** eye.
3. Ask where light falls in relation to red line (Figure 18, 19, 20).
4. Neutralization of deviation. Position vertical prism bar in front of left eye in frontal plane position to neutralize deviation. Note diopters required to neutralize the deviation.

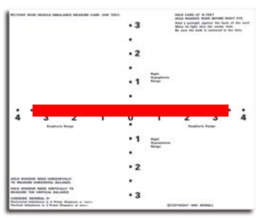

Figure 18. Orthophoria. Red line is seen straight through the light.

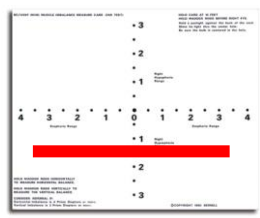

Figure 19. Hyperdeviation. Red line is seen below the light.

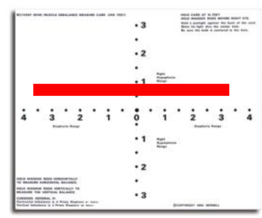

Figure 20. Hypodeviation. Red line is seen above the light.

iii. Consider referral – near phoria cards.
  1. Horizontal balance - Esophoria 5 diopters or more and Exophoria 10 diopters or more
  2. Vertical balance - 2 diopters or more

iv. Consider referral – far phoria cards.
  1. Horizontal imbalance - 4 diopters or more
  2. Vertical imbalance - 2 diopters or more

(5) Three Step Test (Park's test or Bielschowksy's head tilt test). Identification of isolated cranial nerve palsies in patients with the diagnosis of cyclovertical strabismus. Variation of cover-uncover test.

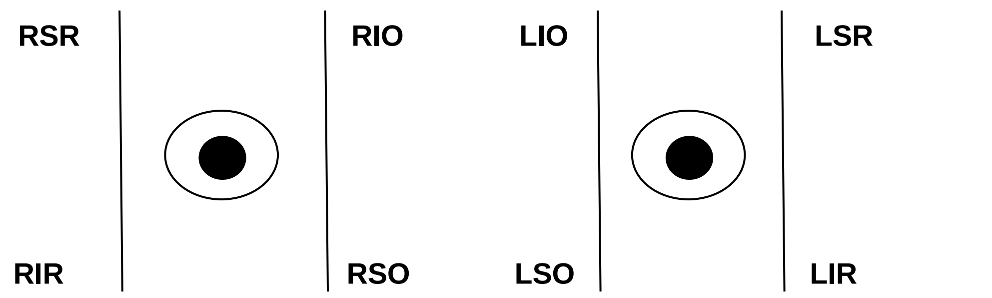

Figure 21. Examiner's view of the patient. Eight cyclovertical muscles are placed in the action field of each respective muscle.[2]

c. Graphical Method[2] (Figure 21).
  i. Step 1: Perform the cover test in primary gaze and determine if there is a right or left hyperdeviation.

1. If there is a right hyperdeviation, circle the two depressors of the right eye (RIR, RSO) and the two elevators of the left eye (LIO, LSR).

ii. Step 2: Perform versions in right and left gaze only. Turn head right 30°test left gaze. Turn head left 30°test right gaze. Determine if the hyper increases in right or left gaze.
   1. If the hyperdeviation increases in left gaze, circle the muscles on the patient's left in each eye (RIO, RSO, LSR, LIR).

iii. Step 3: Perform right and left head tilt. Determine if the hyper increases with head tilt to the patient's right or left shoulder.
   1. If the hyperdeviation increases with head tilt to the right shoulder, circle the muscles that correspond to head tilt to the patient's right in each eye (RSR, RSO, LIO, LIR).

iv. Diagnosis. Muscle circled 3 times is the affected muscle. This example – right superior oblique.

v. Sensitivity of 3-step test in diagnosis of unilateral superior oblique palsy[17]. 70 patients with strabismus who had evidence of superior oblique atrophy on MRI underwent ocular motility testing.
   1. 70% of patients with superior oblique atrophy fulfilled the entire three-step test.
   2. 28% of patients fulfilled 2 of the 3 steps.
   3. 2% of patients fulfilled 1 of the 3 steps.

vi. Sensitivity of 3-step test in diagnosis of bilateral superior oblique palsy[18]. 25 patients diagnosed bilateral SO palsy.
   1. 3-step test had a sensitivity of 24%.
   2. Reversal of hypertropia had a sensitivity of 60%. Reversal of the hypertropia from straight-ahead gaze to any of the other 8 diagnostic positions of gaze.

(5) Double Maddox Rod Test. Tests for cyclotorsion in the clinical setting.

a. Double Maddox rod test procedure.
   i. Trial frame with double Maddox rod oriented at 90°.
   ii. A red lens is placed in front of the right eye and white lens in front of the left eye.
   iii. The subject is asked to align parallel the left beam of light with the right beam of light.
   iv. The amount of torsion is measured from the left trial lens.

b. Sensitivity of double Maddox rod test. Double Maddox rod test is subjective and prone to administrator and subjective error.[19] Normal subjects have more difficulty properly aligning the double Maddox rod as distance between images is increased. Recruitment of the oblique muscles during attempted vertical fusion leads to subjective torsion.

(E) **Differentiating causes of vertical diplopia** – misinterpretation of gravity signal (skew deviation) or paretic eye muscle (trochlear nerve palsy).

(1) Physiological basis of upright-supine test. The utricles detect change in head orientation and skew deviation is caused be a disruption of the utriculo-ocular reflex. When changing from an upright to supine position, the orientation of the utricles changes from earth-horizontal to earth-vertical. In supine, the orientation of the utricle with respect to gravity leads to a saturation or reduction in the overall afferent activities of the untriculo-ocular reflex leads to a reduction of torsion and vertical

misalignment in skew deviation. With a unilateral trochlear nerve palsy, the utriculo-ocular pathway remains intact. Therefore, change in head orientation does not affect vertical and torsional ocular alignment.

(2) Upright-Supine Test.[10] Clinical bedside test used to differentiate skew deviation from trochear nerve palsy.

   a. Patient positioned in the upright position, measure vertical misalignment of the eyes with the alternate cover test and a prism bar and ocular torsion with the double Maddox rod test.
   b. Patient positioned in the supine position, measure vertical misalignment of the eyes with the alternate cover test and a prism bar and ocular torsion with the double Maddox rod test.
   c. Compare measurements obtained in the upright versus supine position.

(3) Tested 10 patients with skew deviation, 14 patients with unilateral peripheral trochlear nerve palsy, and 12 normal participates.[10] Measurements obtained in the supine position compared to the upright position.

   a. Vertical misalignment was decreased by 74% in skew deviation, increased by 5% in trochlear nerve palsy, and increased by 6% in normal.
   b. Torsion was decreased by 83% in skew deviation, by 2% in trochlear nerve palsy, and by 6% in normal subjects.

(4) Sensitivity of specificity of Upright-Supine Test.[10]

   a. In patients with skew deviation, sensitivity of 76%.
   b. In patients with trochlear nerve palsy, specificity of 100%.

(5) Decision making algorithm for vertical strabismus (Figure 22).[10]

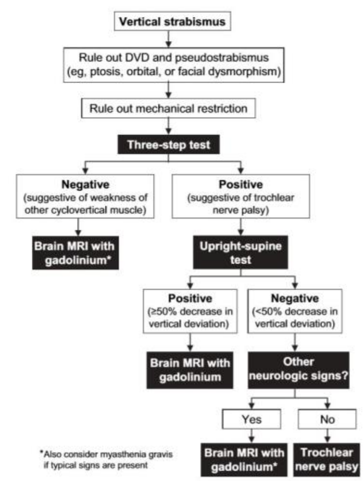

Figure 22. Decision making algorithm for vertical strabismus.

(VI) Case study – Impact of Binocular Misalignment on Vestibular Rehabilitation Outcome in a Patient with Unilateral Vestibular Hypofunction – an Interdisciplinary Approach.

(VII) Case study - Measurement and Management of Binocular Misalignment in Central Vestibular Dysfunction – an Interdisciplinary Approach.

(V) The Process of Differential Diagnosis – Measurement of Subjective Perception of Binocular Visual Signals to Estimate Relative Binocular Alignment – the Vertical Alignment Nulling and Torsional Alignment Nulling Tests.

This case discusses a novel method of oculomotor alignment using the Vertical Alignment Nulling (VAN) and Torsional Alignment Nulling (TAN) tests (**Figure 23**). VAN and TAN are non-invasive, behavioral measures of ocular alignment using a computer tablet, colored lenses, and touch screen software.[20,21]

(A) Perfect alignment (B) Right eye above left eye (C) Right eye

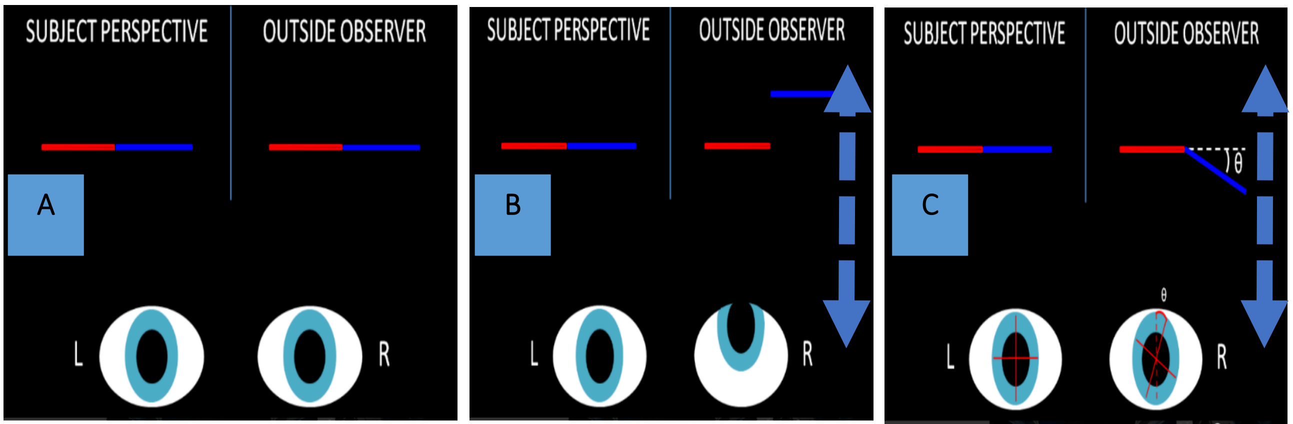

Figure 23. Examples of ocular misalignments inferred by VAN and TAN results. The subject repositions the moving blue line until it appears in line with the stationary red line, thereby positioning each line on the center of each retina. Binocular misalignment is inferred from the relative positioning of the lines at the end of each trial. (A) If the subject has perfect binocular alignment, then the lines will be perfectly aligned at the end of the trial. (B) If the subject sets the blue line above the red line during VAN, we infer that the right eye is elevated above the left eye. (C) If the subject orients the blue line CW relative to the red line in TAN, we infer that the right eye extorted relative to the left eye.

(A) In utriculo-ocular pathology, Skew and Torsion misalignments are head position dependent—they reduce moving from upright to supine.
(B) In trochlear nerve/oculomotor muscle pathology - there is little or no change in the torsional or vertical deviation between the 2 head positions.[22]

(C) Therefore, the Sit to Supine test can help resolve if signs of OTR are vestibular, oculomotor, or central.
(D) Utriculo-ocular pathway causes of skew reduce when positioned supine based on altered otolith signal input[7,10]

(VIII) Case study – Vertical Alignment Nulling and Torsional Alignment Nulling Test in Dysfunction of the Otolith Pathway. The case will discuss the development and validation of this new, portable technology and present results comparing VAN/TAN scores in upright versus supine across different patient populations.

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