Hypertropia:

Vertical strabismus describes a vertical misalignment of the eyes. The misalignment is typically labeled by the higher, or hypertropic, eye, but it can also be labeled by the lower, or hypotropic, eye. Some advocate for labeling based on whether the deviated eye manifests as a hypertropia or hypotropia. Depending on which eye is fixing, a hypertropia of one eye is the same as a hypotropia of the fellow eye. If the degree of deviation in all fields of gaze is the same, it is classified as comitant;

if it behaves differently in different fields of gaze, it is classified as incomitant.

Etiology: Vertical misalignments of the eyes typically result from dysfunction of the vertical recti muscles (inferior and superior rectus) or the oblique muscles (inferior oblique and superior oblique). More rarely, they are caused by abnormal positioning of the horizontal rectus muscles. Muscle dysfunction may result from paresis, restriction, overaction, muscle malpositioning, and dysinnervation.

Due to Muscle Restriction or Overaction:

- **Thyroid Eye Disease**: Leads to enlargement of the extraocular muscles and restrictive strabismus. The inferior rectus is most frequently affected, followed by the medial rectus muscle. Associated findings include increased intraocular pressure when looking away from the restriction, proptosis, lid retraction, compressive optic nerve dysfunction, conjunctival hyperemia, chemosis, and corneal exposure(exposure keratopathy).
- **Brown Syndrome**: Poor movement of the superior oblique tendon through the trochlea leads to limited elevation of the eye in adduction, often with associated exotropia in up-gaze.
    - Congenital causes include inelasticity of the SO muscle-tendon complex, pseudo-Brown's syndrome due to inferior orbital adhesions, and inferior displacement of the lateral rectus.
    - Iatrogenic causes include surgery for glaucoma, oculoplastics, strabismus, or ENT.
    - Inflammatory causes include rheumatoid arthritis and systemic lupus erythematosus.
    - Other causes include trauma and nasal sinus infection.

- **Inferior Oblique Overaction**: Over-elevation of the eye in adduction. If primary and bilateral, it gives rise to a Y-pattern with divergence in up-gaze; if secondary, it may give rise to a V-pattern with convergence in down-gaze. The head tilt test is negative if it is primary. It is frequently bilateral and associated with horizontal strabismus.
- **Superior Oblique Overaction**: Vertical deviation increases on adduction of the affected eye. It frequently coexists with underaction of the contralateral inferior rectus and intermittent exotropia. Can give rise to acquired Brown's syndrome due to SO contracture. A vertical deviation in the primary position is more frequently associated with unilateral or asymmetric SO paresis, and bilateral cases often present with a lambda-pattern (λ).

Orbital Causes of Vertical Restriction:

- Orbital wall (blowout) fracture with entrapment, orbital mass, and orbital or extraocular muscle inflammation can lead to vertical strabismus.

Due to Weak Muscle:

- **Unilateral Superior Oblique Paresis**: Causes hypertropia that increases on adduction and with ipsilateral head tilt. It is the most common cause of an isolated vertical deviation. Features include mild extorsion, compensatory head tilt to the contralateral side, sometimes associated with facial asymmetry, and large vertical fusional amplitudes when congenital.
- **Bilateral Superior Oblique Paresis**: Causes left hypertropia in right gaze and left tilt, right hypertropia in left gaze and right tilt. Larger extorsion than unilateral paresis; esotropia increases in down-gaze.
- **Monocular Elevation Deficit Syndrome (MEDS)**: Involves a limitation of upward gaze with hypotropia that is similar in adduction and abduction. Causes include restriction of the inferior rectus muscle, deficient innervation of elevator muscles, or a combination of both. Clinical features vary based on the underlying cause.

Dissociated Vertical Deviation: An upward drift of one eye when binocular fusion is interrupted without a compensatory downward shift of the fellow eye. Associated features include abduction and extorsion. Increased vertical deviation on head tilt to the ipsilateral side. Most frequently both eyes are affected, although it may be asymmetrical. Any cause leading to disruption of normal binocular development can be at its origin.

Signs and Symptoms:

- **Suppression**: Typically happens when the deviation starts before 6 years of age.
- **Diplopia**: Occurs when the deviation is acquired after significant maturation of the visual system (7 to 8 years of age).
- **Confusion**: Two images are perceived in the same location due to misalignment of retinal correspondence points on the fovea.

Diagnosis:

- **Sensorimotor Testing**: Determine the degree of misalignment for primary, horizontal, vertical gazes, and in head tilt. Torsion can be tested with the double Maddox rod test. Haploscopic testing can evaluate the ability to fuse in the setting of torsion. Worth 4 dot and Bagolini lenses can evaluate for suppression.
- **Differentiating between Paresis and Restriction of the Antagonist**:
    - **Forced Duction Test**: Evaluates for evidence of restriction or laxity.
    - **Saccadic Eye Movements**: Normal saccadic movements until the restrictive amplitude is achieved in restriction cases, where it stops abruptly. Palsy cases show decreased saccadic velocity and force generation.
    - **Intraocular Pressure**: Restrictions may lead to increased IOPs when the eye moves against the restriction.
    - **Lid Fissure**: Restrictions may cause narrowing of the lid fissure, while a paresis causes widening

Three Step Test for Cyclovertical Muscle Palsy

1. **Determine which eye is hypertropic**:
    - The impacted muscle will be a depressor of the higher eye (inferior rectus or superior oblique) or an elevator of the lower eye (superior rectus or inferior oblique).
2. **Determine in which horizontal gaze the hypertropia is worse**:
    - If worse in left gaze, the oblique muscles in the right eye or the vertical recti in the left eye are affected.
    - If worse in right gaze, the oblique muscles in the left eye or the vertical recti in the right eye are affected.

3. **Determine in which head tilt the deviation is worse**:
    - If worse in right tilt, the right eye intorters (superior oblique and superior rectus) or the left eye extorters (inferior oblique and inferior rectus) are affected.
    - If worse in left tilt, the left eye intorters (superior oblique and superior rectus) or the right eye extorters (inferior oblique and inferior rectus) are affected.

Note: The three-step test is not diagnostic when more than one muscle is affected or if there is a restrictive cause. This includes conditions such as:

- Paresis of more than one vertical muscle
- Contracture of the vertical recti
- Previous vertical muscle surgery
- Skew deviation
- Myasthenia gravis
- Dissociated vertical deviation
- Small vertical deviations associated with horizontal strabismus

Cranial Nerve 4 Palsy (Superior oblique palsy)

Dysfunction of the fourth cranial nerve (trochlear nerve), which innervates the superior oblique muscle (SOM), is one cause of paralytic strabismus. The SOM has different (primary, secondary, and tertiary) actions dependent on mechanical position of the eye. In the primary position, the primary action of the superior oblique muscle is intorsion. In adduction, the superior oblique is primarily a depressor. In abducted gaze, the SOM acts to intort the eye and abducts the eye. Thus, a trochlear nerve palsy causes an ipsilateral higher eye (i.e., hypertropia) and excyclotorsion (the affected eye deviates upward and rotates outward). Patients may report vertical and/or torsional diplopia that is usually worse on downgaze and gaze away from the affected side.

Etiology

Isolated 4th Nerve Palsy

Congenital Trochlear nerve palsy is a common cause of congenital cranial nerve (CN) palsy. Patients with congenital CN IV palsies may compensate for diplopia with variable

head positioning; chin-down head posture is seen in bilateral CN IV palsy and contralateral head tilt is typically seen in unilateral CN IV palsy. Later in life, these patients may experience decompensation of their previously well controlled CN IV palsy from the gradual loss of fusional amplitudes that occurs with aging or after illness or other stress event. Congenital CN IV palsies can have very large hypertropias in the primary position (greater than 10 prism diopters) despite the lack of diplopia or only intermittent diplopia symptoms. These large vertical fusional ranges characteristic of congenital cases.

Trauma CN IV has the longest intracranial course and is vulnerable to damage, even with relatively mild trauma. For trauma-induced trochlear palsy, patients typically report symptoms immediately after injury. Bilateral involvement is rare in non-traumatic cases but is relatively more frequent after trauma (crossed, dorsal exit). Careful examination is necessary in traumatic cases as the CN IV palsies can by asymmetric if bilateral and can be masked or become apparent after strabismus surgery for a presumed unilateral CN IV palsy. Computed tomography (CT) scan is generally the first line imaging study in trauma but is often normal.

Microvascular disease Microvascular disease can involve CN IV and usually in older patients with cardiovascular risk factors. Sudden onset, of a painless, neurologically isolated CN IV without a history of head trauma or congenital CN IV palsy in a patient with risk factors for small vessel disease implies an ischemic etiology.

Idiopathic Idiopathic cases may improve or completely resolve over a matter of weeks.

Non-isolated 4th Nerve Palsy

Trochlear nerve palsy can also occur as part of a broader syndrome related to causes like trauma, neoplasm, infection, and inflammation. These etiologies are further categorized based on the anatomic location of involvement (midbrain, subarachnoid space, cavernous sinus, orbit). Increased intracranial pressure has also been known to cause CN 4.

Diagnosis

History

Fourth cranial nerve palsies can affect patients of any age or gender. They can present with vertical diplopia, torsional diplopia, head tilt, and ipsilateral hypertropia. Determining the onset, severity, and chronicity of symptoms can be vital in delineating between the various etiologies of a CN 4 palsy. Furthermore, careful history including

associated symptoms and other past medical history can help distinguish a CN 4 palsy from other items on the differential.

Physical Exam

Examiners should consider obtaining the following: visual acuity, motility evaluation, binocular function and stereopsis, strabismus measurements at near, distance, and in the cardinal positions of gaze, and evaluation of ocular structures in the anterior and posterior segments.

Signs

Ipsilateral hypertropia and excyclotorsion are frequently seen due to the superior oblique's function of intorsion and depression the eye. Patients can also develop a compensatory head tilt in the direction away from the affected muscle.

Symptoms

Patients can present with binocular, vertical or torsional diplopia. The superior oblique causes eye depression in adducted gaze. This can explain the worsening of a patient's diplopia when they attempt to visualize objects in primary position, especially in down-gaze. Patients with mild or long-standing disease may have blurred vision, difficulty focusing and dizziness instead of diplopia.

Unilateral CN IV palsy:

Diagnosis is made via the Parks-Bielschowsky three-step test. Ductional testing may be normal however or only show mild depression deficit in adduction with trochlear nerve palsies. The three questions to ask in evaluation of the CN IV palsy are as follows:

1. Which is the hypertropic eye?
    - There are eight possible muscles that could cause a hypertropia -- the bilateral superior recti, inferior recti, superior obliques and inferior obliques. Determining the hypertropic eye reduces the potentially involved muscles to four. These include the ipsilateral depressors - the superior oblique and inferior rectus or the contralateral elevators - the superior rectus and inferior oblique. For example, with a right hypertropia, the potentially involved muscles include the right superior oblique, right inferior rectus, left inferior oblique and left superior rectus.
2. Does the hypertropia worsen in left or right gaze?

- Determining if the hypertropia is worse in left or right gaze helps eliminate two of the possibly affected muscles. The SOM has action that varies depending on the angle between the muscle plane and the visual axis. When the eye is adducted, the muscle plane and the visual axis align and the primary action is as a depressor. When the eye is abducted the visual axis and the muscle plane become more perpendicular and the SOM function is mostly intorsion. So, in a patient with right hypertropia that worsens in left gaze, this suggests either right superior oblique or a left superior rectus involvement.

3. Does the hypertropia worsen in left or right head tilt? (Bielschowsky head tilt test)

    - Determining if there worsening of the hypertropia in left or right head tilt can identify the involved muscle from the remaining two choices following steps 1 and 2 of the three-step-test. If the hypertropia is worse in ipsilateral tilt this implicates the ipsilateral superior oblique as the intorsional ability of the superior oblique is weakened. In this head position, the ipsilateral superior rectus will compensate for the weak intorsion of the ipsilateral superior oblique but will elevate the eye and further worsen the hypertropia. Patients may develop a compensatory head tilt to the contralateral side to reduce their diplopia. In a patient with hypertropia that worsens in left gaze and right head tilt is most compatible with a right superior oblique palsy.

2. Double Maddox Rod

    - In fourth nerve palsy the Double Maddox rod should demonstrate unilateral excyclotorsion.
    - Skew deviation may demonstrate bilateral torsion or incyclotorsion, both of which are inconsistent with fourth nerve palsy.
    - Bilateral CN IV palsy may have large degree of bilateral excylotorsion (e.g., > 10 degrees) on the Double Maddox rod test

3. Fundus Photography

    - Could demonstrate that the fundus of the affected eye is excyclotorted.
    - Skew deviation may display incyclotorsion of the affected eye or bilateral torsion.

- Bilateral CN IV palsy might show bilateral excyclotorsion.

Bilateral CN IV Palsy

Features suggestive of a bilateral fourth nerve palsy include:

1. Alternating hypertropia on horizontal gaze or tilt
2. Positive Bielschowsky head tilt test to either shoulder
3. Large degree of excyclotorsion (> 10 degrees)
4. Absent or small hypertropia in primary gaze
5. Underaction of both superior obliques on duction testing
6. A V-pattern esotropia of greater than 25 prism diopters