ASSESSMENT OF THE EXTRAOCULAR MUSCLES

AUTHOR

Thomas Salmon: Northeastern State University, USA

PEER REVIEWER

Scott Steinman: Southern California College of Optometry, USA

THIS CHAPTER INCLUDES A REVIEW OF:

- Definitions
- Testing Methods
- Neuromuscular Anomalies (Infranuclear)
- Evaluation

INTRODUCTION

Ocular motility is concerned with how the eyes move. By evaluating the various types of eye movements such as saccades, pursuits and vestibular and optokinetic movements, we can gain information about the ocular motility system. It is the object of regard that determines the type of movement exerted by the eyes. These movements include the following:

- Saccade - the ability to fixate and change fixation
- Pursuit – the ability to follow a target
- Vestibular – the ability to fixate a target while the head position changes
- Optokinetic – the ability to involuntarily fixate on an object that is moving relative to the head.

DEFINITIONS

Concomitancy/Comitant: angle of deviation is the same in all positions of gaze.

Nonconcomitancy/Incomitancy: angle of deviation changes in different fields of gaze.

Paralysis/Paralytic/palsy: nerve damage is complete and no innervation flows to the affected eye muscle.

Paresis/paretic: disruption of innervation is partial, not total. The extent of the paresis may vary from mild to severe.

TESTING METHODS

TOOLS FOR TESTING	**Observation during history:** One of the most important tools used for assessing ocular motility is simple observation during history. Observing the patient walking and looking around the room allows you to look for signs of strabismus, head position in tilts, tips or turns. Head position helps compensate for muscle dysfunction. Anatomical structure can fool you into thinking a strabismus exists. Check the bridge width on children and observe for facial asymmetry. **Clues regarding onset:** Recent diplopia, noncomitancy and abnormal head posture help to distinguish between acquired and congenital problems. Is a new problem more likely to cause diplopia? Which would be more noncomitant? When does head position change? Usually, newer problems are more likely to cause diplopia and be incomitant while the head posture is an adaptation that is generally associated with older defects.
TESTING OF VERSIONS AND DUCTIONS	**Broad H test:** This test is done binocularly. If the deviation measured is comitant, then the strabismus is nonparetic. If the deviation measured is incomitant, suspect one or more paretic EOMS. If you test both eyes, you can have the patient inform you of any diplopia they might be aware of and using a penlight for testing the corneal reflex can also give you clues about the alignment. If over or under actions are noted, retest monocularly. The monocular test identifies which eye has the paretic muscle. If binocular test is normal, monocular testing is not necessary. See fig. 10-9 Further testing helps identify incomitancy: **Maddox Rod test:** This test can be used to evaluate a heterophoria (phoria) as well as the comitancy in various fields of gaze. Prism can be used to align the vertical red line with the light in order to measure the direction and size of the phoria. Be aware that glasses may induce prism that may not be present normally. **Red lens test:** This test is a subjective test used to determine the presence of deviation in the nine positions of gaze. With a red lens over one eye, ask the patient to identify if any separation is observed between the white and red light in the nine positions of gaze. **Parks 3-step test:** This test evaluates vertical deviations in order to isolate the paretic muscle. **Step 1:** in primary gaze which eye is hypertropic. **Step 2:** in which gaze, right or left does the hypertropia increase **Step 3:** when the head is tilted right or left, which has the greatest degree of hypertropia

NEUROMUSCULAR ANOMALIES (INFRANUCLEAR)

Neuromuscular anomalies lead to eye fatigue and discomfort; compensating head position can occur.
Symptoms of diplopia would be present early on, but with time improve, as the visual system compensates.

6TH NERVE PALSY	This nerve is easily damaged due to the length and course of the nerve. Palsy results in the inability to abduct the eye, resulting in esotropia in primary gaze. To compensate, the face is turned towards the affected side in an effort to decrease diplopia. This is the most prevalent, acquired, noncomitant deviation. It is rarely congenital. In young patients consider trauma as a possible cause. In older patients, 6th nerve palsies often develop due to ischemia, related to hypertension or diabetes. Palsies presented in people under the age of 40 may have developed due to multiple sclerosis (MS). Other causes that may be considered include; nystagmus blocking, thyroid myopathy, myasthenia gravis, orbital inflammatory pseudotumor and orbital blowout trauma.
DUANE'S SYNDROME	Duane's syndrome is characterised by a retracted globe and narrowed fissure when adduction is attempted and an inability to abduct the eye. It is the left eye that is most often affected. 15-20% of cases are bilateral and are more prevalent in females compared with men with a 4:1 ratio. **Etiology of Duane's Syndrome:** The traditional explanation is that the lateral rectus is fibrotic. An alternative theory is the mis-wiring of the nerves supplying the lateral rectus. Innervation goes to both medial rectus (MR) and the lateral rectus (LR) by the 3rd nerve causing contraction of both muscles, which results in retraction of the globe, partial limitation of adduction and narrowing of the palpebral fissure. **Classification of Duane's Syndrome** **Duane's Type I** • Very limited abduction in the affected eye • Lids may widen on abduction • Slightly restricted adduction • Upshoot or downshoot on adduction • Retraction of the globe and narrowing of the palpebral fissure on adduction • Binocular in primary gaze usually, but may be esotropic • Observe for head turn in direction of the affected eye. • Limited binocularity **Duane's type II** • Very limited adduction • Relatively normal abduction • Retraction on abduction with lid narrowing • Also known as Inverse Duane **Duane's type III** • Limited or absent adduction and abduction • Retraction of the globe and narrowed fissures on adduction • Rarest of the 3 types **Differential diagnosis:** MR palsy, LR palsy, thyroid myopathy, spasm of near reflex, esotropia **Treatment:** Find cause and rule out other causes, give prescription (Rx), monitor, provide vision therapy (VT), prescribe prism and/or patching. Most patients learn to move their head. With strabismus, the suppression helps prevent diplopia. Consider surgery if cosmesis is a concern.

MÖBIUS SYNDROME:	Möbius syndrome is characterised by a LR palsy that is bilateral. There is also lack of facial musculature, a decreased bulk of one side of the tongue, mental retardation, congenital heart defects, limb and chest deformities, hearing, speech and swallowing problems.
4TH NERVE PALSY	The thinness and position of the CN IV makes it vulnerable to injury. Traumatic closed head injury from a frontal blow is a very common cause. Even minor head injury can be enough to cause nerve damage. A 4th nerve palsy causes paresis of the superior oblique (SO). It is the most common cause of vertical deviation (90%), where the hypertropia increases in inferonasal gaze. Torsional diplopia occurring in other positions increases temporally. The parks 3-step test will show this. If the lesion is in the nucleus or located prior to decussation, the defect is contralateral and if the lesion is at the site of decussation the result is bilateral paresis. If the lesion occurs post decussation, the result is ipsilateral paresis. The third nerve may often be involved resulting in a down and outward turn of the eye that cannot be moved into the correct field of gaze to assess the 4th nerve. Watch for torsion of the eye to see if the 4th nerve is intact. **Position of eye and head:** The affected eye is hypertropic in primary gaze. There may be an excyclotorsion and a small esotropia in the primary gaze. There is no depression when the eye is adducted. The chin is depressed allowing for up gaze. The head is tilted away from the affected side towards the opposite shoulder. The falling eye sign is observed when the yoke muscle in the contralateral eye is the inferior rectus; so when fixating in the field of gaze of the affected eye, the other eye may drop due to increased innervation to the inferior rectus. **Causes of CN IV palsy:** • Trauma: 35% • Congenital: 60% • Vascular infarct from diabetes mellitus (DM) or hypertension (HTN) • Idiopathic • MS • Tumour • Aneurysm 21% of CN IV nerve palsies are bilateral and caused by head trauma. The patient has diplopia and tilts the head. In order to see bilateral involvement, an alternate cover test in right and left gaze should be performed. **Differential diagnosis:** • Myasthenia gravis - characterised by fatigue in up gaze • Thyroid myopathy • Orbital inflammatory pseudotumour **Treatment:** Find cause; prescribe prism, VT, spot patching, monitor.

BROWN'S SYNDROME	Brown's syndrome is characterised by: • An inability to elevate the eye when adducted • Chin elevation • Minimal vertical deviation in primary gaze – poor parks 3-step test • Normal to slight limitation of elevation in primary gaze and abduction • Often binocular in primary gaze, • Monocular but can be binocular (10%) • Congenital or sometimes due to trauma • Widening of the lids on adduction • Divergence in up gaze - V pattern exotropia • Caused by mechanical anomaly of the SO; the tendon sheath is short and fixed at the pulley and will become a barrier as the eye tries to adduct. The globe will slip under the stretched sheath and you will hear a click as the patient tries to elevate the eye during adduction. **Differential diagnosis:** IO paresis – a forced duction test will rule out this. **Treatment:** Find cause, give Rx, monitor, VT, prescribe prism, spot patching. Surgery not recommended unless there is a significant vertical strabismus. Many people with Brown's syndrome have normal binocular vision. **Remember:** Lesions in the cortex and supranuclear connections can cause conjugate dysfunction in saccades, pursuits or vestibular movements. Lesions in the midbrain that interfere with the gaze centres and internuclear connections can result in the inability to produce conjugate eye movements. It is only at the motor nuclei level that damage will be present in one eye or the other.
3RD NERVE PALSY	The CN III is the thickest of all the cranial nerves. Lesions may not interfere with the entire function of the nerve. Compressive and traumatic lesions of less impact may disrupt a portion of the nerve. Pupil function may be spared. The location of the lesion along the nerve pathway determines which functions are lost. Total block prior to where the nerve splits leads to ipsilateral paralysis of the medial rectus (MR), inferior rectus (IR), inferior oblique (IO), superior oblique (SR), the levator, pupillary sphincter and ciliary muscle. The eye is dilated, unable to focus, has ptosis and a divergent strabismus. The eye is turned down and out. Incomplete paresis results in combinations of these signs and symptoms. Superior branch lesions will affect the superior rectus and levator; the inferior branch lesions will affect the inferior rectus, medial rectus and motor root to the ciliary ganglion. Lesions in the oculomotor complex can affect some nuclei and save others. Many variations are possible. **Etiology of CN III nerve palsy:** **Pupil sparing:** A pupil sparing palsy is often due to vascular causes such as DM or Cavernous sinus syndrome where deeper parts of the nerve are affected. **Pupil involved:** A pupil-involved palsy indicates a more compressive lesion such as an aneurysm, tumour, trauma and rarely herpes zoster. When the pupil is blown and fixed there is internal ophthalmoplegia, which involves the pupil and all of the CN III. **Appearance of the patient:** The eye will be turned down and out with only temporal movement and slight inferior movement as only the 6th and 4th nerve are operating. The lid may be closed, the pupil blown, and diplopia present.

3RD NERVE PALSY	**Differential diagnosis:** • Myasthenia Gravis • Thyroid myopathy • Chronic progressive external ophthalmoplegia • Orbital pseudo tumour • Midbrain lesion • Severe hypertension **Treatment:** Find the cause, immediate CT and/or MRI if: 1. Pupil involved 2. Pupil is spared but the patient is less than 50 years of age, no history of DM, no improvement after 2 to 3 months, other nerves affected Give prescription (Rx), Fresnel prisms, possible spot patching, VT for calisthenics and sensorimotor fusion training.
SR PALSY	A palsy of the superior rectus presents with a hypotropia and ptosis of the involved eye. Can you explain why? The ptosis occurs because these nerve fibers travel together in the superior branch of CN III. Hypotropia increases in abduction with the head tilted towards the involved side as seen with the Parks 3-step test. Can you explain why it increases with abduction? Hypotropia increases in abduction due to the secondary action of the superior rectus in abduction, which is due to the insertion of this EOM. The patient cannot elevate the eye during abduction; there may be extorsion of the eye if acquired as an adult. The chin is usually elevated to keep the eyes in down gaze. An isolated superior rectus palsy is very rare and is usually congenital. **Differential diagnosis:** Mechanical causes for limitation: • **Thyroid myopathy:** Thyroid myopathy will present with proptosis, inflamed EOMs and lid retraction. With thyroid problems, the IR is most often affected and then the MR> SR> LR. Obliques are rarely affected by thyroid disorders. • **Trauma:** Trauma such as a blow-out fracture may trap the IR and lead to poor elevation • **Ocular myasthenia:** In cases of superior oblique palsy, ocular myasthenia is an inhibitional palsy of the contralateral antagonist. **Treatment:** Find cause, give RX, VT, prism if needed, spot patching.
MR PALSY	• A palsy of the medial rectus presents with an exotropia of the involved eye, an inability to adduct the eye, the face turns away from the affected side to place the affected eye in temporal gaze. • An isolated MR is extremely rare • This palsy may occur from a blowout fracture **Differential diagnosis:** • INO (Internuclear Ophthalmoplegia): where the affected eye cannot adduct to look horizontally. INO is caused by lesions in the medial longitudinal fasciculus (MLF). A unilateral INO suggests an infarct in the branch of the basilar artery. A bilateral INO (BINO) is a red flag for MS. • Duane's syndrome, type II • Thyroid myopathy • Ocular Myasthenia **Treatment:** Find cause, give Rx, VT, prisms, spot patching.

IR PALSY	A palsy of the inferior rectus presents with: • Hypertropia of the involved eye • The hyper deviation increases in abduction and with head tilt to the opposite side • Intorsion, if acquired as an adult • Inability to depress the eye during abduction • The chin is depressed slightly • It is the most frequent muscle to become fibrotic **Differential diagnosis:** • Trauma • Thyroid myopathy • Ocular myasthenia **Treatment:** Find cause; give Rx, VT, prisms, spot patching and monitor.
IO PALSY	A palsy of the inferior oblique presents with: • Hypotropia of the involved eye with some intorsion. • An inability to elevate the eye during adduction • The chin is elevated, with the head tilted towards the involved side and the face turned towards the non-involved side • Hypotropia increases in adduction and on head tilt towards the unaffected side on the parks 3-step test • An A-pattern exotropia is seen in down gaze • IO is the least likely muscle to be affected and if it is, it is usually congenital.

EVALUATION

ANGLE LAMBDA (OR KAPPA)	It is the angle between the pupillary axis and the line of sight. Normally this measures +5 degrees; the plus indicating that the line of sight is nasal to the pupillary axis, a minus indicating that the line of sight is temporal. The average position of the corneal reflex will be 0.4mm nasal to the centre. A difference between the two angle Lambdas suggests eccentric fixation.
HIRSCHBERG TEST	With both eyes unoccluded and the patient fixating a penlight at 50cm, compare the positions of the corneal reflexes in both eyes, under binocular conditions. Then compare these measurements with the position of the reflex under monocular conditions. This test reveals the presence or absence of a strabismus and is very useful in children, infants and those with poor cooperation. 1 mm of deviation indicates 22 prism dioptres of strabismic deviation. Compare the reflex difference starting from the position of the fixating eye. Do not start from the centre of the pupil. **Example:** If the reflex is 0.5mm nasal on the right eye and 1mm temporal on the left eye, how much of a difference is between the two? The total difference is 1.5mm leading to 30 prism dioptres of exotropia. These reflexes can be recorded with photos to help in assessment.
KRIMSKY REFLEX PRISM AND BRUCKNER TESTS	Similar to the Hirshberg, this test uses prism placed before the fixating eye to equalize the reflexes. The Bruckner test uses an ophthalmoscope where the practitioner sits 50cm to 1m from the patient having them look directly at the light. The practitioner focuses on the anterior surface and watches both eyes. The colour and symmetry of the red fundus reflex is evaluated. The whiter, brighter eye is the one that is turning.
UNILATERAL AND ALTERNATE COVER TESTS	The unilateral cover test reveals the presence of strabismus. The presence and magnitude of a heterophoria is measured using the alternating cover test. Prism is used to neutralize the movement. 7 prism dioptres of deviation is estimated for every 1 mm of movement.

RECOMMENDED READING

Reading Chapter 10 I in Benjamin, W. Borish's Clinical Refraction. WB Saunders, Philadelphia. 2006.

BIBLIOGRAPHY

Ciuffreda KJ and Tannen B. **Eye Movement Basics for the Clinician**. Mosby, St. Louis, 1995.

Hart W. **Adler's Physiology of the Eye, 9th Ed**. Mosby Yearbook, St. Louis. 1992.

Steinman et al. **Foundations of Binocular Vision**. McGraw-Hill, New York, 2000.

Regan D. **Binocular Vision (Vol 9 in Vision and Visual Dysfunction, 1991)**.

Reading RW. **Binocular Vision**. Butterworth Publishers, Woburn, MA, 1983.

Schwartz S**. Visual Perception - 2nd Edition**. Appleton & Lange, Stamford, CT, 1999.

Griffin JF. **Binocular Anomalies - Diagnosis and Vision Therapy, 3rd Edition**, Butterworth-Heineman, 1995.

Kaufmann, PL. **Adler's Physiology of the Eye, 10th Ed.** Mosby, St. Louis, 2003.

Moses, RA. **Adler's Physiology of the Eye, 8th Ed**. Mosby Yearbook, St. Louis. 1987.

Kandel. **Essentials of Neural Science and Behavior**, Appleton & Lange, 1995.