LEARNING OBJECTIVES

1. The attendee will be familiar with the common presentations of non-organic ocular motor disturbance.

2. The attendee will be able to distinguish these psychogenic manifestations from similar organic neuro-ophthalmic syndromes using specific examination techniques.

3. The attendee will be able to distinguish pharmacologic mydriasis from parasympathetic pupillary palsy using clinical features and pharmacologic testing.

CME QUESTIONS

1. Voluntary nystagmus is usually accompanied by which of the following:
   a. Vertical gaze palsy
   b. Ptosis
   c. Convergence
   d. Pupillary dilation

2. Reported causes of convergence spasm include which of the following:
   a. Myasthenia
   b. Botulism
   c. Wernicke’s encephalopathy
   d. Radiation therapy

3. Conditions that may mimic non-organic convergence spasm include which of these:
   a. Ocular neuromyotonia
   b. Posterior fossa tumor
   c. Myasthenia
   d. Midbrain stroke
   e. All of the above

4. The most helpful feature for distinguishing Adie tonic pupil from pharmacologic mydriasis is:
   a. Light-near dissociation
   b. Segmental sphincter palsy
   c. Conjunctival blanching
   d. Response to weak pilocarpine

KEYWORDS

1. Convergence spasm
2. Spasm of the near triad
3. Pseudomyopia
4. Voluntary nystagmus
5. Pharmacologic mydriasis

INTRODUCTION

Somatoform neuro-ophthalmic disorders most commonly affect afferent visual function. When eye movements, eyes or pupils are involved, the diagnosis may be challenging. In many such cases the variable nature of the abnormality in question suggests the correct diagnosis but great care must be taken because certain organic disorders may be similarly intermittent. For each of the non-organic syndromes described below, one or more “look-alike” conditions are also described, including features that help distinguish them from the non-physiologic variety.

EYE MOVEMENTS

Gaze palsies

Non-organic conjugate (horizontal or vertical) gaze palsies are most often seen as an incidental finding in a patient with other non-organic visual disturbance(s) or in a patient who is especially anxious about the examination. In the latter, adopting a reassuring and non-threatening manner and moving the fixation target further back from the patient may be sufficient to produce normal versions. In other cases, observing random eye movements during other parts of the visit (like during the history) will reassure the examiner that eye movements are in fact full.

The Lookalike: some eye movement disorders produce a dissociation between saccadic and pursuit movements. This can create the impression of inconsistency and thus be mistaken for a non-organic eye movement disorder. The key is to recognize that the variability in range of motion is related to the type of eye movement tested. In most such cases there is more limitation to saccades (as tested by voluntary re-fixations) than to pursuit movements (a common feature in patients with Progressive Supranuclear Palsy). Similar dissociation of eye movements is seen in...
some cases of internuclear ophthalmoplegia in which adduction is preserved for convergence movements but not for conjugate lateral gaze and in cases of ocular motor apraxia (congenital or acquired) in which reflex movements accomplish what cannot be done with voluntary gaze.

Convergence spasm
Most convergence spasm occurs as a component of the near triad, accompanied by pupillary constriction and accommodation. The large majority of such patients do not have an underlying organic cause, although cases due to brain disease have been described [See Table 1]. When convergence spasm does occur as a manifestation of brain disease it is most often due to midbrain dysfunction and is accompanied by other signs of midbrain disturbance such as up-gaze palsy and poorly reactive pupils with or without light-near dissociation.

The determination that intermittent esotropia is due to convergence spasm is usually based on the observation of miosis while the eyes are converged. The associated induced myopia can often be observed during retinoscopy. In addition, the limitation of abduction induced by convergence spasm often disappears under monocular viewing. Treatment is often challenging [See Table 2].

Look-alike: variable esotropia may occur with myasthenia but the lateral rectus is not a favorite target in this disease and is therefore uncommon. Ocular neuromyotonia, when it affects the medial rectus, also produces variable esotropia. In such examples of this uncommon disorder, failure of medial rectus relaxation following adduction causes persistent esotropia upon returning to primary position, worse on contralateral gaze, lasting from seconds to minutes. The trick to diagnosing this condition is recognizing that the variable esotropia is not random or voluntary but precipitated by gaze in a particular direction. Diagnosis may be more challenging in cases in which contractions also occur spontaneously and in those with a refractory period after contraction. A helpful clue for diagnosing this syndrome is recalling that it usually occurs after radiation therapy for a skull base tumor. Dramatic response to carbamazepine is characteristic.

Look-alike: accommodative spasm occasionally occurs in isolation, not accompanied by miosis and convergence. The most common form of isolated accommodative spasm, also termed pseudomyopia, is peripheral, i.e. due to ciliary spasm as from inflammation, corneal abrasion or trauma. Uncommonly, pseudomyopia is due to central nervous system disease, usually severe head injury. Accommodative spasm due to head trauma usually affects young adults (perhaps because of their more robust accommodative amplitudes) and can persist for years. The basis for such spasm is unclear, most likely due to disinhibition of putative brain stem accommodation centers. Cases are usually managed by supplying the manifest (non-cycloplegic) refraction (or at least part of it) or with cycloplegic drops and near glasses.

Look-alike: acute acquired comitant esotropia (AACE) usually arises from temporary occlusion of the visual axis that disrupts fusion of a pre-existing esophoria. Occasional cases are due to brain disease, the most worrisome being a posterior fossa tumor.

NYSTAGMUS
Some normal subjects can voluntarily induce saccadic oscillations, termed “voluntary nystagmus”. The ability to produce such movements is found in 5 – 8% of the population and sometimes runs in families. These eye movements are conjugate and are usually confined to the horizontal plane with frequencies ranging from 3 – 42 Hz and amplitudes from 0.5 to 35 degrees. Eye movement recordings in such individuals indicate that these ocular oscillations are more similar to ocular flutter and opsoclonus than to nystagmus and the alternative term “psychogenic flutter” has been proposed. In most individuals with this talent, saccadic oscillations are accompanied by convergence and fluttering of the eyelids and the presence of these features should suggest the correct diagnosis.

EYE LID CLOSURE
Voluntary ptosis does not usually occur in isolation but is found more often in the setting of other non-organic visual/ocular symptoms following trauma or other inciting incident. While this condition may simulate levator weakness, more often it takes the form of orbicularis spasm; at times the patient presents a picture of both levator weakness and orbicularis spasm which is especially helpful in confirming its non-organic basis. It is difficult to sustain voluntary ptosis in a consistent fashion; the twitchiness of the apparently ptotic lid is a helpful feature but must be distinguished from the lid twitches of myasthenia and from aberrant regeneration involving the levator. It is difficult to sustain voluntary ptosis and upgaze at the same time and so observation of lid position on upward gaze is a helpful examination technique.

The Lookalike: patients with Essential Blepharospasm, as in other forms of dystonia, often have tricks they can use to improve their symptoms. For example, some patients find that opening their mouth helps them keep their eyes open. Such oddities of basal ganglia function may give the impression of a non-organic movement disorder. In addition, patients with blepharospasm may be worse under scrutiny with bright lights. Thus, with “casual conversation”, as when engaged in the history-taking, they may not exhibit much in the way of unwanted eyelid closure.

PUPILLARY DILATION
Pharmacologic instillation with a cholinergic agent produces pupillary dilation that may be suspected based on the very large degree of dilation and a history of access to a mydriatic agent. Trauma to the pupillary sphincter and anterior chamber inflammation are ruled out by biomicroscopy, the demonstration of normal ocular motility rules out a third nerve palsy. The non-organic basis for this form of mydriasis is typically confirmed by its lesser response to pilocarpine compared to the fellow eye.
The Lookalike: an acute post-ganglionic parasympathetic palsy (Adie's pupil) produces a similarly dilated and poorly reactive pupil to light. Characteristic light-near dissociation and super-sensitivity to weak cholinergic agents are later developments and therefore not helpful for diagnosis in the acute setting. In contrast to pharmacologic mydriasis, an Adie pupil at any stage should show a strong miotic response to 1-2% pilocarpine. The most helpful feature for distinguishing pharmacologic mydriasis from an Adie's pupil is the segmental nature of the sphincter palsy in the latter.

Table 1. Organic Causes of Convergence Spasm

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<tr>
<th>Condition</th>
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<td>Diphenylhydantoin intoxication</td>
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<td>Wernicke's encephalopathy</td>
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<td>Head trauma</td>
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<td>Chiari I malformation</td>
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<td>Syphilis</td>
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<td>Labyrinthine lesions</td>
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Table 2. Reported Treatment Options for Spasm of the Near Triad

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<th>Option</th>
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<td>Cycloplegic eye drops and reading glasses</td>
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<td>Anti-anxiety medication</td>
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<td>Placebo eye drops</td>
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<td>Counseling</td>
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<td>Glasses with opaque inner third</td>
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<td>Amytal interview</td>
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CME ANSWERS

1. c
2. c
3. e
4. b

REFERENCES