Movement Disorders
CLINICAL PRACTICE
EDITORIAL COMMENTARY

# Eyelid Nystagmus and Other Involuntary Movements of the Upper Lids; What's in a Name?

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In this issue, Ordás and colleagues[1] report a case of "eyelid nystagmus" in a 75-year-old woman presenting with 5 years progressive gait disorder on a background of hypertension. Other than the eyelid nystagmus, the only abnormal features were limb hyperreflexia and a "slow gait with decreased step height" interpreted as a "higher-level gait disorder". Eye movements were normal and in particular there was no ocular nystagmus, and therefore, this was isolated eyelid nystagmus. Brain magnetic resonance imaging showed moderately extensive small vessel white matter change in the cerebral hemispheres and pons. The authors propose first, that the nature of the abnormal lid movement was myorhythmia or slow tremor, and second, that the cause was the ischemic change in the pons.

The case report highlights several unresolved issues on the nomenclature and classification of involuntary movements of the upper lid. It is timely then to consider what is the phenomenon of eyelid nystagmus and where does it fit within the classification of involuntary lid movements. Is it really "nystagmus" or an alternative type of movement disorder that demands an alternative descriptive term? It should be noted that the literature descriptions generally refer to involvement of the upper lid with either no involvement, or at least no description, of the lower lid. An accurate description and classification of the phenomenology is critical to the determination of causality and mechanism.

The upper lid elevates in upgaze and lowers in downgaze during saccades and pursuit to protect the cornea while maintaining full vision. It moves at a velocity and amplitude that generally matches that of the underlying globe.[2] The central caudal nucleus (CCN) of the oculomotor nerve in the midbrain innervates the levator palpebrae and the superior rectus muscles bilaterally. Therefore, the two muscles are yoked and contract in unison. This function requires coordination of eye and lid activity. The CCN maintains a tonic level of activity during eye opening that increases with upward eye movements and decreases with downward eye movements.[2,3] Normally, upper lid movements result from antagonistic activity of levator palpebrae and orbicularis oculi, which reciprocally inhibit each other.[4] This coordination is mediated by the superior colliculus (SC). The SC projects to the supraoculomotor area directly overlying the CCN as well as to the facial nuclei and is inhibited by the pars reticulata of the substantia nigra. A group of midbrain neurons, the M-group, send projections to the CCN and receive excitatory input from the rostral interstitial nucleus of the median longitudinal fasciculus—which generates vertical saccades—and the SC during upgaze, and inhibitory input from the interstitial nucleus of Cajal and nucleus of the posterior commissure during downgaze. In this way, upper lid elevation and depression is modulated in concert with vertical eye movements.[3] Involuntary eyelid movements, such as eyelid nystagmus, are likely to result from inappropriate or disordered excitation or inhibition of levator palpebrae or orbicularis oculi or both.[5]

A number of abnormal upper lid abnormal movements have been described, with terms that include lid nystagmus, lid fluttering, lid hopping, lid tremor, lid myoclonus, lid myokymia, lid fasciculations, and, by Ordás and colleagues[1] in this issue, lid myorhythmia. Rarely, such movements are confined to the upper lid (ie, isolated), whereas some are associated with simultaneous ocular movements (eg, most cases of lid nystagmus, eyelid myoclonus), and others with contractions of orbicularis oculi (eg, blepharoclonus and blepharospasm). Figure 1 depicts a suggested algorithm for classifying involuntary upper lid movements based largely on phenomenology.

Historically, there have been three types of eyelid nystagmus reported. These are described and discussed by Ordás et al,[1] in this issue and well summarized and referenced in their Table 1.[1] The most common is that associated with vertical ocular nystagmus, such that with upbeat nystagmus there is an upper lid movement (upward jerk) synchronous with the globe, but with an exaggerated excursion. The second type is associated with

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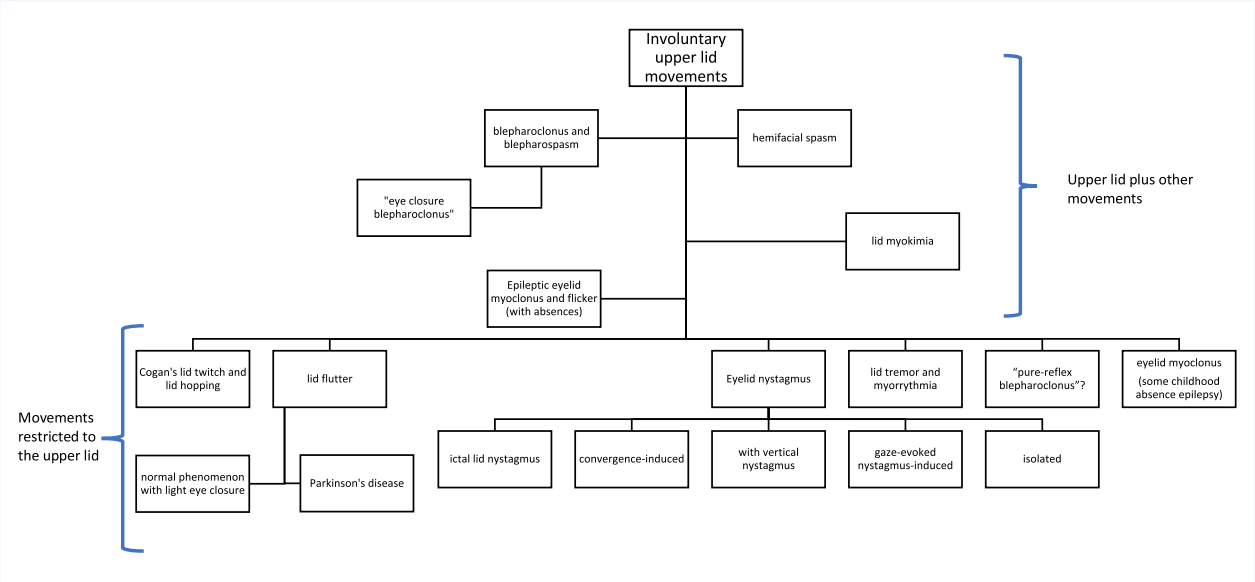


**FIG. 1.** A proposed classification algorithm for involuntary upper lid movements. These have been separated into those that are accompanied by additional movements beyond the upper lid (eg, inferior orbicularis) and those that are not.

gaze-evoked horizontal nystagmus so that there is an upward jerk of the lid jerks simultaneously with the fast phase of the horizontal gaze-evoked nystagmus. The third type is convergence evoked lid nystagmus, whereby active convergence induces upward jerks of the lids in the absence of ocular vertical nystagmus. Interestingly, this can be maximal with the lids lightly or incompletely closed.[6] In each of these three types of lid nystagmus there is a rapid upward movement of the lid followed by a slower downward restoration of the lid position. Therefore, there is a distinct similarity to ocular jerk nystagmus and so the term "lid nystagmus" is apt. The neural basis for the striking and perplexing dissociation or "uncoupling" of motion of the upper lid from that of the superior rectus muscle that characterizes eyelid nystagmus, remains speculative and not fully explained, and especially given their common innervation via a shared midbrain nucleus and superior division of the third cranial nerve.

Ordás et al[1] propose that their patient exhibited "a form of slow tremor or a form of myorhythmia" with electromyography of the levator palpebrae exhibiting a semi-rhythmic activity of 1–2 Hz. Their accompanying video does indeed show slow semi-rhythmic elevation and lowering of the upper lid without a clear jerk component. Vial and Hallett[7] in their review of myorhythmia highlight the very variable description of this phenomenon in the literature and propose that it be defined as a slow periodic rhythmic or semi-rhythmic involuntary movement of some part of the body and express some doubt that it should be regarded as a tremor. Therefore, it would seem that the appropriate description of the upper lid movement in the patient reported by Ordás et al[1] is lid myorhythmia rather than lid nystagmus.

Eyelid tremor has been infrequently reported.[8,9] Jungehülsing and Ploner[8] described a patient with a right paramedian thalamic infarct and rhythmic jerks of the lids with a frequency of 7 Hz on voluntary eye closure and abolished with forced eye closure. This was termed "tremor" by the authors, although no video was provided with the publication. Ueno and colleagues[9] reported a patient with anti-Caspr2 antibody-related encephalitis presenting with parkinsonism and "eyelid tremor" at a frequency of 1–2 Hz. However, the accompanying video appears to show repetitive semi-rhythmic blinking rather than typical tremor, perhaps better classified as a type of blepharoclonus. Therefore, isolated lid tremor as a classification or entity has yet to be convincingly established.

Lid myokymia (incorrectly also referred to in the lay literature as lid fasciculations) is observed as continuous, multicentric, small amplitude contractions of the orbicularis oculi muscles, especially inferiorly. It is a benign, usually unilateral, self-limiting phenomenon experienced by otherwise healthy individuals and associated with fatigue, anxiety and depression, excessive caffeine intake, and some drugs. More persistent eyelid myokymia can be encountered in multiple sclerosis, Guillain Barre syndrome, brainstem tumors, and in the setting of coronavirus disease 2019 infection.[10] Eyelid myoclonus can occur as a generalized epileptic phenomenon usually with absences and precipitated by eye closure in the light.[11] There is jerking of the eyelids immediately after eye-closure. The eyelid movement resembles rapid blinking rather than lid nystagmus and the globes deviate upward. This differs from the slight myoclonic flickering of eyelids accompanying typical childhood absences where there is no movement of the globes. Similarly, ictal blinks can be an uncommon epileptic feature.[12] In contrast, ictal lid nystagmus is characterized by a rapid upward lid movement and slow drift downward without movement of the eyes or orbicularis oculi involvement and, therefore, can be classified as one of the forms of isolated lid nystagmus.[12]

Repetitive upper lid movements occur in blepharoclonus and blepharospasm, but usually in concert with contractions of the

orbicularis oculi. Blepharospasm is characterized by repetitive tonic eye closure from involuntary contraction of orbicularis oculi muscles. Blepharoclonus is distinguished by brief, repetitive, clonic contractions of orbicularis oculi.[13] Another lid movement is eye-closure blepharoclonus induced by voluntary light eye closure. Jacome[14] described eye-closure blepharoclonus as tremor or myoclonus of the lids, sometimes associated with orbicularis oculi contractions, in several patients with a range of neurological disorders. In others, the abnormal movements were restricted to the upper lid and termed "pure-reflex blepharoclonus." Eye-closure blepharoclonus remains to be confirmed as a distinct entity. It may be, in this author's view, an enhanced form of the normal phenomenon of eyelid flutter on light eyelid closure. Healthy individuals when asked to lightly close the eyes will develop an irregular fast fluttering of the upper lid, abolished with firm eye closure.[4] The author has observed this eyelid flutter to be present in the majority of healthy people (unpublished observations). Eyelid flutter on light eye-closure has been also highlighted in Parkinson's disease[15,16] suggesting that there is a disinhibition or increased activation of a normal phenomenon, and presumably mediated by an altered substantia nigra pars reticulata–superior colliculus pathway.[17]

Hemifacial spasm begins in the orbicularis oculi muscle before ultimately spreading to the lower facial muscles, with sometimes prominent involvement of the upper lid where there is high frequency twitching of the pre-septal and pre-tarsal component of the muscle.[3] However, the spasms are rarely confined to the upper lid with the lower lid also involved, and of course, as the name implies, the spasms are unilateral. Lid hopping, describes the fluttering of a ptotic eyelid, especially during horizontal eye movements, and mostly encountered in myasthenia gravis.[18–20] A related phenomenon in myasthenia gravis is the excessive conjugate upward twitch of the upper lid when the eyes return to primary position after downgaze, first described by Cogan[18] and commonly referred to as "Cogan's lid twitch."

In conclusion, involuntary movements of the upper lid can be classified according to the nature of the movement (phenomenology), the precipitating factors (eg, convergence), and the associated oculomotor (eg, nystagmus) and neurological features (eg, orbicularis contraction, facial contractions, and parkinsonism) (Fig. 1). The field requires a greater number of detailed clinical observations with consistent nomenclature, allied to electrophysiological, neuroimaging, and neuropathological studies to better understand the underlying mechanisms and aberrant neural underpinnings of these involuntary lid movements, including lid nystagmus. Such advances would facilitate a more complete classification than that suggested here.■

## Author Roles

(1) Research project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript: A. Writing of the First Draft, B. Review and Critique.

T.A.: 3A, 3B.

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## References

1. Ordás CM, Querejeta-Coma A, Yupanqui-Guerra L. Isolated eyelid nystagmus: case report and Nosological considerations. *Mov Disord Clin Pract* 2023. https://doi.org/10.1002/mdc3.13743
2. Hamedani AG, Gold DR. Eyelid dysfunction in neurodegenerative, neurogenetic, and neurometabolic disease. *Front Neurol* 2017;8:329.
3. Rucker JC. Normal and abnormal lid function. *Handbook of Clinical Neurology [Internet]*. Amsterdam, The Netherlands: Elsevier; 2011 [cited 2023 May 24]:403–424 Available from: https://linkinghub.elsevier.com/retrieve/pii/B9780444529039000212.
4. Schmidtke K, Büttner-Ennever JA. Nervous control of eyelid function: a review of clinical, experimental and pathological data. *Brain* 1992;115(1):227–247.
5. Aramideh M, De Visser BWO, Koelman JHTM, Bour LJ, Devriese PP, Speelman JD. Clinical and electromyographic features of levator palpebrae superioris muscle dysfunction in involuntary eyelid closure. *Mov Disord* 1994;9(4):395–402.
6. Sanders MD, Hoyt WF, Daroff RB. Lid nystagmus evoked by ocular convergence: an ocular electromyographic study. *J Neurol Neurosurg Psychiatry* 1968 Aug;31(4):368–371.
7. Vial F, Hallett M. Myorhythmia. In: Testa MC, Haubenberger D, eds. *Tremors [Internet]*. 1st ed. New York: Oxford University Press; 2022 [cited 2023 May 26]:191–195 Available from: https://academic.oup.com/book/43955/chapter/369588389.
8. Jungehulsing GJ. Eyelid tremor in a patient with a unilateral paramedian thalamic lesion. *J Neurol Neurosurg Psychiatry* 2003;74(3):356–358.
9. Ueno S, Hirano M, Sakamoto H, Kusunoki S, Nakamura Y. Eyelid tremor in a patient with anti-Caspr2 antibody-related encephalitis. *Case Rep Neurol* 2014;6(2):222–225.
10. Khan HA, Shahzad MA, Jahangir S, et al. Eyelid myokymia—a presumed manifestation of coronavirus disease 2019 (COVID-19). *SN Compr Clin Med* 2022;4(1):29.
11. Giannakodimos S, Panayiotopoulos CP. Eyelid Myoclonia with absences in adults: a clinical and video-EEG study. *Epilepsia* 1996;37(1):36–44.
12. Pyatka N, Gajera P, Fernandez-Bacavaca G, Lhatoo SD, Shaikh AG. Ictal lid movements: blinks and lid saccades. *Neuro-Ophthalmol* 2021;45(5):301–308.
13. Obeso JA, Artieda J, Marsden CD. Stretch reflex blepharospasm. *Neurology* 1985;35(9):1378–1380.

14. Jacome DE. Synkinetic blepharoclonus. *J Neuro-Ophthalmol Off J North Am Neuro-Ophthalmol Soc* 2000;20(4):276–284.

15. Loeffler JD. Motor abnormalities of the eyelids in Parkinson's disease: electromyographic observations. *Arch Ophthalmol* 1966; 76(2):178.

16. Beltre N, Feldman M, Marmol S, Shpiner D, Luca C, Moore H, et al. Blepharoclonus as a potential novel clinical marker in Parkinson's disease (P3-11.009). *Sunday, April 23 [Internet]*. Philadelphia: Lippincott Williams & Wilkins; 2023 [cited 2023 May 24]:4702. Available from: http://www.neurology.org/lookup/. https://doi.org/10.1212/WNL.0000000000204169.

17. Basso MA, Powers AS, Evinger C. An explanation for reflex blink hyperexcitability in Parkinson's disease. *I Superior Colliculus J Neurosci* 1996;16(22):7308–7317.

18. Cogan DG. Myasthenia gravis: a review of the disease and a description of lid twitch as a characteristic sign. *Arch Ophthalmol* 1965;74(2):217.

19. Weinberg DA, Lesser RL, Vollmer TL. Ocular myasthenia: a protean disorder. *Surv Ophthalmol* 1994;39(3):169–210.

20. Ragge NK, Hoyt WF. Midbrain myasthenia: fatigable ptosis, "lid twitch" sign, and ophthalmoparesis from a dorsal midbrain glioma. *Neurology* 1992;42(4):917–919.