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Abnormal head oscillations in neuro-ophthalmology and neuro-otology

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Abstract

Purpose—This paper describes the clinical phenomenology and contemporary pathophysiology of concurrent oscillations of the eyes and the head that are present in neurological conditions with diverse etiologies.

Recent findings—One classic example is spasmodic nuttans in which the eye oscillations are the primary pathology while head nodding is thought to be an operant conditional response that suppresses the eye oscillations to facilitate clear vision. The second example is a combination of head tremor and inadequate compensatory eye movements due to vestibular hypofunction leading to the illusion of pendular nystagmus – hence the condition is called pseudonystagmus. Cerebellar degeneration in ataxia-telangiectasia or synchronized spontaneous discharge from the inferior olive and cerebellum in the syndrome of ocular palatal tremor with dystonia are examples of eye and head oscillations due to the impairment in cerebellar outflow. Infantile nystagmus syndrome also presents with eye and head oscillations, but here the relationship between the two types of oscillations differs among patients and is still unclear.

Summary—The clinical features and putative pathophysiology of diverse disorders of the eyes and head that produce unwanted oscillations are reviewed and some analogies between eye and head oscillations are suggested. Key clinical pearls that are essential to separate these disorders at the bedside are also emphasized.

Keywords

tremor; nystagmus; opsoclonus; myoclonus; cerebellum; brainstem

Introduction

Novel stimuli appearing in the visual field triggers reorienting movements of the eyes and the head. These synchronized eye and head movements rely on common neural correlates. For example, the superior colliculus responsible for initiating eye saccades also generates rapid head movements via tectocollic pathway. Neurons in the midbrain and pontine

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reticular formation generate a burst of discharge for both the eyes and the head. Neural integrators in the midbrain have eye-head neurons that guarantee precise and well-coordinated eye and head movements. Finally, the vestibulo-ocular reflex (VOR) generates compensatory eye movements in response to head movements to ensure steady fixation of targets that are stationary in the environment. Such closely tied and well-synchronized eye and head movements assure clear vision to targets of interest whether they are moving or still but are at risk of malfunction in any of its components. Such abnormalities can be classified into the following broad categories: (1) Eye movements as the primary abnormality with the seemingly abnormal head movements actually being compensatory. (2) Head movements as the primary disorder with seemingly abnormal eye oscillations being compensatory. (3) Abnormal eye and head oscillations due to a shared pathophysiology. (4) Eye and head oscillations occurring independently. Here we review these categories of head oscillations seen in patients with various neuro-otological or neuro-ophthalmological conditions.

Primary eyes oscillations and consequent head oscillations

Spasmus nutans

Spasmus nutans occurs in early childhood and consists of eye and head oscillations; the latter is often called “head nodding”. This classic condition was described in the late 19th century and it was later shown that the eye and head oscillations have different sources [1, 2]. Gresty and colleagues (1976) first discovered a link between the eye and head oscillations in one patient with spasmus nutans [3]. In this case report the head and eye oscillations were recorded and the authors found two characteristic features. The ocular oscillations were sinusoidal, disconjugate, had a frequency of 11 Hz and up to 2 degrees in amplitude. The head was largely stable during eye oscillations, but there were intermittent epochs of head nodding during which the abnormal eye oscillations disappeared. The head movements, however, were of a much lower frequency and larger amplitude than the eyes oscillations,. The authors proposed that head nodding in spasmus nutans is not a pathological phenomenon but a compensatory mechanism that leads to improved vision by turning off the central mechanisms that drive the abnormal eye oscillations. [3]. In this formulation the effect of the head movements is indirect in the sense that their dynamic characteristics such as amplitude and timing do not mirror those of the eyes. These features of the eye and head oscillations were subsequently confirmed in several subsequent studies [4].

Primary head oscillations and consequent eye oscillations

Pseudonystagmus

The vestibulo-ocular reflex (VOR) facilitates clear vision during locomotion by producing eye movements equal and opposite to head movements in order to stabilize gaze. Vestibular hypofunction leads to suboptimal ocular motor compensation and a false sensation of movement of the visual surround known as oscillopsia. The combination of a constant head tremor and vestibular hypofunction can produce a persistent distressing oscillopsia in a syndrome called pseudonystagmus [5-8]. The constellation of head tremor and vestibular hypofunction has been reported in patients with tremor due to multiple sclerosis, and to

immunosuppressive medications in patients with renal transplantation who also have antibiotic ototoxicity, or in essential head tremor [5-8].

Figure 1 depicts an example of the eye and head movements during intended gaze holding in a patient with pseudonystagmus. Time-locked pendular oscillations of the head and eyes (actually gaze (or eye position in space) as the eye position in the orbit is actually still) are evident in the horizontal and vertical planes. The frequency of eye oscillations (3.9 ± 0.7 Hz) is comparable to head oscillations. The mean amplitudes of the horizontal head and eye oscillations were also comparable; $2.1 \pm 0.8^\circ$ and $1.6 \pm 0.8^\circ$ respectively. The phase locked (180° out of phase) eye and head oscillations, suggested that the unstable gaze was secondary to a hypoactive VOR in the presence of a continuous head oscillation.

Head impulse testing further confirmed VOR hypofunction. Figure 2A,B illustrates the reduced peak velocity of the compensatory eye movement responses (black traces) during multiple trials of high-velocity, horizontal and vertical head impulses (gray traces). Also, the peak eye velocity lagged behind the peak head velocity. Figure 2C,D compares the horizontal and vertical head velocity with the corresponding compensatory horizontal and vertical eye-in-orbit velocity (signs inverted) during head impulses. The delayed compensatory eye movement responses resulted in an elliptical trace. The smaller peak velocity of the compensatory eye movement is reflected in an ellipse with the long axis steeper and directed away from the equality line.

In sum, diminished compensatory eye movements, secondary to vestibular hypofunction, in patients with head tremor can cause oscillopsia, mimicking the symptomatic presentation of a pendular nystagmus. Unless the head is stabilized, especially during ophthalmoscopy or slit-lamp examination, the pseudonystagmus falsely gives an impression of a pendular eye nystagmus, a disorder that has a quite different pathogenesis and options for treatment. This example of pseudonystagmus emphasizes the importance of stabilizing the head during ophthalmoscopy, particularly in patients with head tremor, in order to avoid the false diagnosis of a pathological eye nystagmus.

Concurrent eye and head oscillations due to the same pathophysiology

Ataxia-telangiectasia

Ataxia-telangiectasia (A-T) is an autosomal recessive degenerative neurological disorder that progressively affects the movements of the eyes, head, limbs, and trunk. Its well-known features include abnormalities of eye movements including defective gaze-holding, trunk ataxia, gait instability, and adventitious appendicular and head movements [9-12].

Quantitative analysis of head movements in A-T revealed at least two different subtypes. One type has a relatively low frequency, in the range of 0.5 to 1 Hz, but large amplitude, between 3-6 degrees. Figure 3 depicts the large jerky and irregular head oscillations in an A-T patient. The second subtype is sinusoidal and has a relatively high frequency ranging between 3-5 Hz but smaller amplitude ranging between 0.5-1 degrees. Such a sinusoidal waveform can be superimposed on the jerky waveform (blue arrows, Figure 3).

Quantitative analysis of the jerky waveforms of the head movements suggests possible mechanisms and also analogies with abnormal eye oscillations. The jerky waveform is comprised of a slow drift in head position followed by rapid corrective, saccade-like head movement. The drift velocity is related to the eccentricity of the head on trunk orientation. At one orientation the drift is minimal; this position is called the null orientation. The drift velocity increases as the head-on-trunk orientation deviates farther from the null. The polarity of the drift velocity is opposite on the two sides of the null position, with the slow drift of the head being towards the null. This systematic relationship between drift velocity and head-on-trunk orientation is plotted in Figure 4A. These quantitative features of the head movement instability suggest a dysfunction of a head neural integrator that holds the head in eccentric positions on the torso. This conceptualization draws on an analogy to the eye position neural integrator, which holds eccentric positions of gaze in the orbit. Presumably this abnormality of head holding may be related to the considerable impairment in cerebellar function in subjects with A-T, analogously to their impairment of holding eccentric eye positions in the orbit. [13-16].

Impaired cerebellar feedback to a head neural integrator can explain the head oscillations much as impaired cerebellar feedback has been evoked to explain the underlying difficulty in holding the eye in eccentric positions in the orbit^[17,18]. Figure 3 depicts an example of impaired gaze holding in the A-T patient. The blue traces show right eye position while the grey traces represent the left eye. The eyes are relatively stable in straight-ahead orientation. When the eyes turn to the right, there are relatively slow leftward drifts away from the target. Rapid corrective movements in the opposite direction bringing the eyes back to the target. In contrast, rightward drifts and leftward correction appear when the eyes are turned to the left. The slow-phase velocity of the drifts systematically changes with the eye-in-orbit position; the velocity reduces as the eye-in-orbit position approaches straight-ahead orientation (Figure 4B). These features are hypothesized to reflect suboptimal neural integration due to impaired cerebellar feedback [10-12, 17, 18]. The abnormal oscillations of the eye and head in A-T are an example in which both the eye and head movement disorders share the same pathophysiology.

Ocular palatal tremor

The syndrome of ocular palatal tremor (OPT) is a degenerative neurological condition affecting the inferior olive and the function of the cerebellum. Acquired lesions involving a strategic anatomical location in the Guillian-Mollaret triangle lead to a degenerative hypertrophy of the inferior olive. The Guillian-Mollaret triangle is comprised of fibers connecting the inferior olive to the deep cerebellar nuclei and the fibers from the deep cerebellar nuclei to the inferior olive that traverse the red nucleus and descend to the inferior olive in the central tegmental tract. The typical cause of OPT is an ischemic stroke or hemorrhage affecting structures in the posterior fossa. The hallmark of this condition is a coarse, irregular and usually disconjugate oscillation of the eyes and a tremor of the palate. The coarse and irregular features of these oscillations resemble myoclonus, hence the older name of ocular palatal “myoclonus”, but the pathophysiology suggests this is not a true myoclonic disorder. We recently described the clinical characteristics of a novel variant in

which OPT was associated with dystonia (OPTd) [19]. We discovered head tremor in this variant of OPTd [19].

Figure 5 describes the characteristics of the eye oscillations in OPTd. Eye oscillations are coarse, irregular, disconjugate and are present in all three dimensions (Figure 5). The oscillation frequency in this example is in the typical range between 2-4 Hz (histograms in Figure 5). The removal of visual fixation or head stabilization does not alter the characteristics of these oscillations. Figure 6 depicts an example of head oscillations in an OPTd subject who attempted to hold the head on a stationary target. Detailed examination revealed irregular, coarse, jerky head oscillations mixed with sinusoidal head tremor just as described previously in subjects with idiopathic cervical dystonia [13, 14] or here in A-T. The frequency of horizontal head oscillations ranged between 3-6 Hz (histograms in Figure 6). The head oscillations were present in both the horizontal and vertical directions. The differences in the frequency of the eye and the head oscillations were reflected in their temporal asynchrony. These results suggested that head oscillations were neither compensatory nor causative for the eye oscillations. Likewise the eye oscillations were not compensatory physiological eye movements as would be the usual VOR response to the head oscillations, but they were part of the OPTd phenomenology.

Elegant animal experiments demonstrated that disruption of Guillian-Mollaret triangle leads to degeneration and abnormally increased soma-somatic gap junctions in the inferior olive [20-24]. The electric coupling through these gap junctions leads to increased but abnormal synchronous output from the inferior olive. We suggested that synchronized inferior olive oscillations are then altered by a maladaptive discharge from the cerebellum [25, 26]. The action potentials from the inferior olive neurons travel into climbing fibers and give rise to complex spikes in the Purkinje cells. The branch of climbing fibers also projects to the deep cerebellar nuclei and then to the Purkinje neurons via parallel fibers and the granular cell layer. Hence, the synchronized oscillations from the inferior olive are received twice by the Purkinje neurons and deep cerebellar nuclei, once through the climbing fiber projection and then after a small delay via the parallel fibers and the granular cell layer and Purkinje cell projections. Such redundant and randomized time delayed signals to the Purkinje neurons could cause maladaptive learning and induce an independent cerebellar signal that would distort the output of inferior olive passing through the cerebellum [26]. We have speculated that maladaptive spontaneous cerebellar output, as a result of hyperactivity in the inferior olive and cerebellar pathway, in patients with OPTd leads to the eye, head, and palate oscillations [26].

Independent (or possibly interdependent) eye and head oscillations

Infantile nystagmus syndrome

Head oscillations often accompany the infantile nystagmus syndrome (INS) (previously called the congenital nystagmus syndrome but in fact in many infants the nystagmus does not appear until several months of life) [27-29]. Most investigations revealed 2-3 Hz head oscillations in patients with INS [28-30]. A fundamental question has been: are these head oscillations in some way adaptive to compensate for the eye nystagmus or are they a pathological abnormality in the cephalomotor system much as is the eye nystagmus in INS

is a pathological abnormality in the ocular motor system? A previous study has shown that both are possible [28]. In one case the VOR reflex is turned off and the head then moves opposite to the eyes and cancels the effect of the eye oscillation. In the other case, the VOR is not turned off, and hence the head oscillations are not adaptive but presumably a manifestation of an abnormality in the cephalomotor system. We recently discovered two types of head oscillations in INS patients [31]; one type had 2-3 Hz frequency, consistent with previous observations [28-30]. The second novel subtype had the higher frequency in the range of 5-8 Hz [31].

Figure 7 depicts an example of the two subtypes of head oscillations and corresponding eye nystagmus from an INS patient. The top panel represents horizontal and vertical eye positions depicting primarily horizontal jerk nystagmus with superimposed mild vertical nystagmus giving an oblique movement trajectory. Two discrete types of oscillations are evident in the head position trace. The low-frequency subtype has a predominantly horizontal trajectory, with a minor vertical component. In contrast, the high-frequency subtype, in the 5-8 Hz range, was episodic and had a prominently vertical movement trajectory. The high-frequency head oscillations did not affect the eye nystagmus, and the trajectories of the head oscillations and eye oscillations were asynchronous. The interpretation was that the high-frequency head oscillations were not adaptive but rather arose from another pathological process reflecting reverberations in the cephalomotor system [31]. This interpretation was consistent with a previous proposal that even the low-frequency head oscillations reflect pathological reverberations in the cephalomotor system [28]. Other investigators, on the contrary, proposed that low-frequency head oscillations are the consequence of a central adaptive strategy to minimize nystagmus [29, 30, 32]. Much as the eye oscillations of the infantile nystagmus syndrome remain poorly understood, the same can be said for the head oscillations.

Other causes of head oscillations associated with ophthalmological conditions

Infantile esotropia, nystagmus, and head tremor

The association of infantile esotropia, nystagmus, and head oscillations was reported in three patients [27]. The surgical realignment of infantile esotropia was associated with resolution of head oscillations. The head oscillations were episodic and their mechanism or usefulness unclear. Two of the three reported patients also had a global developmental delay. Such developmental delay might suggest coexistence of two independent conditions common in neurodevelopmental disease – esotropia and head stereotypies.

Conclusions

- Concurrent eye and head oscillations can have a similar phenomenology but wide range of etiologies.
- In patients with spasmus nutans the eye oscillations are the primary pathology while head nodding is an operant conditional response suppressing the eye oscillations to facilitate clear vision. How the nystagmus is suppressed is unknown

- Pseudonystagmus is an example featuring the combination of head tremor and inadequate compensatory eye movements due to vestibular hypofunction causing the illusion of pendular nystagmus if the head is not properly stabilized when examining the eyes, for example, using the ophthalmoscope or a slit lamp.
- Degenerative neurological disorders, such as ataxia-telangiectasia, or the very rare syndrome of ocular palatal tremor associated with dystonia, are examples of eye and head oscillations putatively due to the same pathophysiology.
- Interdependence of eye and head oscillations in infantile nystagmus syndrome is controversial.
- The quantitative characteristics and the pathophysiology of abnormal head oscillations have been relatively little studied and, as our understanding of how the brain controls movement continues to improve, remain wide-open for investigation. In particular, quantitative recording of head movements, using paradigms similar to those used in studying eye movements, are particularly fruitful

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Figure 1.

Example of attempted fixation with and without head restraint in a patient with multiple sclerosis had a head tremor, vestibular hypofunction and a consequent pseudonystagmus. Eye-in-orbit position (movement of the eye in reference to head) and head-in-space position (head movement in reference to space fixed coordinate) are plotted on the y-axis while time is on the x-axis. Panels A,B depict head-in-space position when the patient's head was passively immobilized. Panels C,D illustrate simultaneously measured eye-in-orbit position. (D) Panel depicts vertical upward drifts in the eye-in-orbit position followed by the downward rapid correction, a downbeat nystagmus also shown by this patient. Panels E,F depict head-in-space position when the head was unrestrained and panels G,H depict corresponding eye-in-orbit positions. Simultaneous eye and head pendular oscillations are seen in the head unrestrained condition. Such eye oscillations are not seen in the head was stabilized. Vertical sinusoidal oscillations characterizing pseudonystagmus are superimposed on the downbeat nystagmus in the head unrestrained condition (adopted from reference 6).

Figure 2.

VOR (vestibulo-ocular reflex) responses to head impulses. (A,B) Head velocity during horizontal and vertical head impulses plotted against time. Gray traces represent superimposed head velocities from several trials of head impulses, and corresponding eye velocity is plotted in black. Normally, head rotation transients are accurately tracked by compensatory eye movements of equal amplitude, so that the gray and black traces overlap. Our patient had a reduction and a delay in the peak eye velocity, indicating a diminished VOR response. (C,D) Head velocity plotted against eye velocity. Normal response is characterized by traces falling along the dashed equality line. The elliptical trace suggested a delay in peak eye velocity. Reduced peak velocity of compensatory eye movements during head impulses is represented in the oblique orientation of the long axis of the ellipse relative to the equality line (adopted from reference 6).

Figure 3.

Example of head and eye oscillations in patients with ataxia-telangiectasia. The head movements were recorded as the subject attempted to keep the head to the right, straight-ahead, and to the left. Note the slow drifts and rapid corrections of the head. There are also sinusoidal head oscillations superimposed upon the slow drifts (most prominent during rightward and straight position). The second panel depicts an example of gaze-evoked eye nystagmus from the same patient. The patient attempted to fixate gaze to the right, straight-ahead, or to the left. The nystagmus is characterized by slow phase and rapid corrective quick phase. Slow phases drift the eyes toward the null.

Figure 4.

The slow-phase velocities of the head (A) and the eye (B) oscillations systematically change with head-on-trunk (A) or eye-in-orbit orientations (B). Each symbol depicts the slow-phase head (A) or eye velocity (B) during one cycle of jerky head (A) or eye movement (B). The slow-phase velocity is plotted on the y-axis while corresponding head-on-trunk or eye-in-orbit position is plotted on the x-axis. Positive signs depict rightward direction, while negative is the leftward.

Figure 5.

(A-C) An example of eye oscillations in one patient with OPTD. Horizontal (A), vertical (B), and torsional (C) eye position is plotted on the y-axis and time (in seconds) on the x-axis. A two second epoch of the eye movement recording is demonstrated in this figure. Colored traces depict the right eye and grey the left eye. (E-G) Histograms summarize the distribution of oscillation frequencies. The x-axis depicts the frequency of oscillations (in bins) while the y-axis is a number of cycles in a given frequency bin. Red histogram depicts horizontal oscillation frequency, the blue vertical (F), and the green torsional frequency (G) (reproduced from reference 19).

Figure 6.

(A) An example of head oscillations in one patient with OPTd. Horizontal (red) and vertical (blue) head position plotted on the y-axis while x-axis shows time in seconds. A two-second epoch of head movement is demonstrated. (B,C) Histograms summarize the distribution of head oscillation frequency. The x-axis shows (in bins) the frequency of oscillations while y-axis corresponds to the number of cycles in a given frequency bin. The red histograms depict horizontal oscillation frequency while blue histograms depict vertical (F)

Figure 7.

This is an example of the eye and head oscillations in a patient with INS. Cyan and magenta lines depict horizontal and vertical eye positions plotted on the y-axis and corresponding time on the x-axis. A simultaneous measure of horizontal and vertical head positions are plotted as red and blue lines, respectively. Red arrow depicts high-frequency oscillations while blue arrow depicts low-frequency oscillations (reproduced from reference 31; with permission).