Abnormal Head Position and Head Motion Associated with Nystagmus

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INTRODUCTION

Over the 40 different types of nystagmus presently identified (Dell'Osso, 1984), only a few are associated with oscillations of the head or specific head positions. We will discuss those which occur in congenital nystagmus (CN) latent/manifest latent nystagmus (LMLN), the nystagmus blockage syndrome (NBS), spasmus nutans (SN), periodic alternating nystagmus/periodic alternating gaze deviation (PAN/PAGD), and acquired downbeat nystagmus. We will also discuss descriptions of spontaneous and induced "head nystagmus." Vestibular nystagmus induced by head positions (positional nystagmus) or rapid head movements (positioning nystagmus) will not be discussed.

Whereas abnormal head positions are probably always adaptive and compensatory, head oscillations need not be. In each instance we will attempt to identify what affect the oscillation has upon perception (visual acuity.)

CONGENITAL NYSTAGMUS

Patients with CN may exhibit either head oscillations, a head position preference or both. The head oscillations seen in CN were originally thought to be compensatory in nature (Cogan, 1967). The patient was said to be moving his head equally and oppositely to the CN and was thereby stabilizing the eyes in space. In the absence of good eye movement recordings, this became an attractive hypothesis. Metz, and colleagues (1972) provided support for the compensatory hypothesis by simultaneously recording eye and head movements. However, examination of their data reveals a jerky head movement of approximately 30 degrees supposedly compensating for a pendular CN of about 7 degrees. Patients with CN rarely complain of oscillopsia during either steady fixation or while moving (actively or passively). Based on clinical observations and personal experience in sports, we concluded that the vestibulo-ocular response (VOR) of the CN patient functions normally. Recent studies have confirmed the normal functioning of the VOR in CN patients (Carl, and colleagues, 1985; Gresty, and colleagues, 1985). We further reasoned that for the head oscillation to be completely compensatory, the patient had to simultaneously turn off his VOR (i.e., gain reduced to zero) and duplicate exactly the CN waveform (180 degrees out-of-phase) (Daroff, and colleagues, 1978). Our earliest recordings revealed the extreme complexity and high bandwidth of these waveforms and we concluded that the above-described total compensation was impossible. Moreover, the foveation periods of each waveform precluded the necessity for totally

stabilizing the eyes in space since for most of each cycle the fovea was off-target and visual information suppressed (this inferred from the lack of oscillopsia) (Dell'Osso, 1973; Dell'Osso and Daroff, 1975). Compensation could be realized during relatively stable (as opposed to motionless) foveation periods with head oscillations and VOR suppression.

Gresty, and colleagues (1978), in a study of a single case, showed by accurate eye and head movement recordings that the head oscillation is merely an extension of the CN and that the normal VOR keeps the eyes stable in space during the foveation periods of the CN waveform. The head oscillations in some patients, under certain conditions, emerge as a manifestation of their ocular motor instability (damped by the mass of the head) and do not result in any acuity improvement or decrement. We concluded, as did Gresty and Halmagyi (1981) and, more recently, Carl, and colleagues (1985), that this is the operable mechanism for head oscillation in most CN patients. The observations that head oscillations appear only during intense fixaton attempts and the accompanying increase in the nystagmus, (i.e., while trying to see targets at the limits of the patient's acuity) and are more prevalent in children who are unconcerned about appearance, support this hypothesis.

There have been recent reports of rare CN patients in which the head oscillation was said to be either truly compensatory or to cancel the eye oscillation (Gresty and Halmagyi, 1981; Gresty, and colleagues, 1984; Taylor, 1980). Here, the word "compensatory" is taken to mean a deliberate head movement that is equal and opposite to an ongoing eye movement with the VOR gain decreased to zero (this usage is opposite to the compensatory eye movement produced by a head movement and the normal VOR gain of one). Unfortunately, no recordings were shown in two of these papers and in the third, the patient identified as having CN with head movements that canceled the nystagmus had convergence (180 degrees out-of-phase) nystagmus that was probably spasmus nutans (see below). Furthermore, the example given for compensatory head oscillation was not convincing; it corresponded exactly to what one would record from a CN patient with a normal VOR operating to negate the effects of his spontaneous head oscillation. We have never observed cancellation of CN by active head shaking.

The only way to differentiate true compensatory head movements from spontaneous head movements would be to compare the foveation periods of the CN waveform when the head is still with the same foveation periods measured during head oscillation. If foveation periods are flat (i.e., eyes motionless) when the head is still, the eye-in-head oscillations seen during head motion are VOR movements added to the CN and the resulting gaze (eye-in-space) tracings will also show flat foveation periods. If foveation periods show the same waveform when the head is still as when it is moving, the head oscillation is truly "compensatory" and only the gaze tracings will show flat foveation periods. Thus, recordings of the CN waveforms under both conditions are necessary to prove the existance of compensatory head movement. To date, we have only observed true compensation of CN by head movements in the data from one CN patient (Carl, and colleagues, 1985). As we had originally predicted, this patient suppressed his VOR gain and improvement occurred only during foveation periods. With the head still, the foveation periods were not flat and when head movement occurred, the gaze tracing showed flat foveation periods. Patients who have achieved flat foveation periods without head movement can gain nothing by head shaking and any head motion should be presumed to be due to the CN. Those whose foveation periods are not flat may improve them with head motion and their head shaking might be suspected to be compensatory.

Patients with CN often adopt head turns that place their eyes at a preferred gaze angle where the nystagmus is minimal and the acuity maximal. The nulling of CN occurs when the eyes are deviated to the null angle or the head is turned in the opposite direction; this is the basis for both prism and surgical therapies. Up to about 15 degrees, the head turn equals the null angle but beyond that, the

amount of head turn is less than that required to balance out the null angle (Fujiyama, and colleagues, 1983). While the exact mechanism of CN nulling is unknown, we hypothesize that the unstable ocular motor system has an equilibrium position (the null angle) that divides two regions of instability, each producing runaway eye movements in opposite directions. The push-pull nature of brainstem neural integrator circuitry is consistent with this conceptualization.

The mechanism underlying abnormal head postures in CN patients that are not simple rotations in the horizontal plane is more obscure. Head tilts toward one shoulder or chin-up and chin-down postures raise the possibility that, for non-strabismic patients, a vestibular input is being used since these postures cause a change in static vestibular firing due to the effects of gravity on the otoliths. Such head postures must be tested under the effects of different gravity vectors (e.g., while the body is in different orientations) before this hypothesis can be evaluated against the alternative; that is, the position of ocular motor equilibrium in such patients requires innervation of muscles in other than the horizontal plane.

LATENT/MANIFEST LATENT NYSTAGMUS

Another congenital type of nystagmus, that is not CN, is latent/manifest latent nystagmus (LMLN) (Dell'Osso, and colleagues, 1979). LMLN is a jerk nystagmus with a decreasing-velocity slow phase that beats in the direction of the fixating eye, the other eye being covered (LN) or open but cortically suppressed (MLN). In many cases of LMLN, the amplitude of the oscillation varies monotonically with gaze angle such that as gaze is directed toward the fast phase, the amplitude increases (Alexander's law). Because of this, and the fact that the ocular motor system seems to be unable to prolong foveation when the slow phases are of decreasing velocity, patients may adopt a head turn to increase their acuity. By turning the head toward the fast phase the fixating eye is placed in adduction where the amplitude of the LMLN is minimal. In cases where the patient has alternate fixation, the head turn will depend on the fixating eye, which will be placed in the adducted position. In patients whose LMLN does not vary in accordance with Alexander's law, a head turn that places the fixating eye in abduction may be preferred. In either case, the head turn will minimize the LMLN and maximize the visual acuity by the resulting increase in foveation time. There may be other factors in some LMLN patients (e.g., an angle kappa) that can result in paradoxical head turns that increase the nystagmus amplitude (Dell'Osso, and colleagues, 1979).

The observation that for the LMLN waveform, the initial rapid movement of the fovea off target precludes plastic adaptation of the ocular motor system deserves comment. In CN waveforms, the slow phases start slowly and increase in velocity as the fovea is driven off target. This low-velocity initial movement is apparently countered by a strong fixation reflex that maintains target foveation and we see the results in the long foveation periods of many CN waveforms. The inability of the LMLN patient to similarly distort the slow phases of LMLN suggests that fixation reflexes are ineffective in the presence of high retinal slip velocities.

NYSTAGMUS BLOCKAGE SYNDROME

The nystagmus blockage syndrome (NBS) is the reduction of a CN or conversion of a CN to a low-amplitude MLN by a purposive esotropia (Dell'Osso, and colleagues, 1983). In either instance acuity is improved. As the esotropia increases, the nystagmus decreases and as the fixating eye follows a target moving laterally towards primary position and then into abduction, the esostropia decreases and the nystagmus increases. The fixating adducted eye necessitates a head turn toward that eye for targets that are in primary position. Often an alternating head turn accompanies alternating fixation in these patients such that the adducted eye is fixating primary-position targets. A head turn is not present when the fixating eye is in primary position and the other (suppressed) eye is

esotropic. These head positions mimic those of LMLN patients whose waveforms follow Alexander's law and who are esotropic; this has caused many such patients to be mis-diagnosed as NBS patients. The head turn in the NBS is adopted by the patient to allow fixation of a primary-position target by the adducted eye.

SPASMUS NUTANS

Spasmus nutans (SN) (a condition that has been described by many names: nutatio capitis spastica; gyrospasm; nodding spasm; rotatory spasm; head nodding; and salaam tic) is a benign, self-limiting condition of infants and children that consists of pendular nystagmus (often asymmetric and/or disconjugate) usually associated with head nodding or anomalous head position. Since the characteristics of the nystagmus are described in this volume by Weissman and colleagues, we will confine ourselves to the head movements. They may be inconstant and irregular and may be absent in some patients.

In the first quantitative recording of both the eye and head oscillations in SN, Gresty, and colleagues (1976) showed that the head oscillation canceled the nystagmus, leaving only the normal VOR movements. This cancellation of the nystagmus in SN by head shaking has since been confirmed in other patients (Gresty and Ell, 1981; Gresty and Halmagyi, 1981; Gresty, and colleagues, 1984); in addition we cite the above-mentioned example of cancellation of "CN" by head shaking (Taylor, 1980). The operative mechanism appears to be one of total cancellation of the ongoing nystagmus by intentional head shaking accompanied by the normal VOR. This differs from the head oscillations of CN which are either produced by the instability itself or are compensatory (i.e., used with a suppressed VOR to stabilize the eyes in space only during the foveation periods). Head shaking improves acuity for SN patients and those few CN patients with compensatory head movements; it is unaffected in most CN cases because of the normal VOR. The cancellation effect explains earlier reports of head nodding without nystagmus in SN (Norton and Cogan, 1954). Nothing is known about the relationship between head posturing and either the nystagmus of SN or visual acuity in SN.

PERIODIC ALTERNATING NYSTAGMUS/PERIODIC ALTERNATING GAZE DEVIATION

Periodic alternating nystagmus (PAN) is a jerk nystagmus that may be either acquired or congenital. It changes direction periodically (or aperiodically if CN) and may include neutral periods during which either no nystagmus or pendular nystagmus is present. Periodic alternating gaze deviation (PAGD) is a condition in which the eyes alternately and conjugately deviate to either side. PAGD has been observed in a patient who previously had PAN with the same periodicity; they are assumed to be related (Kennard, and colleagues, 1981).

The mechanism of PAN has been linked to a periodic shifting of the nystagmus neutral region and to the optokinetic-vestibular system (Daroff and Dell'Osso, 1974; Leigh, and colleagues, 1981). In both PAN and PAGD cases, there may be a periodic head turning that is in counterphase with the eye oscillations or deviations. It appears that the purpose of this deliberate slow head turning is to keep the region of minimal PAN straight ahead or, in the case of PAGD, to facilitate fixation of targets in primary position.

ACQUIRED DOWNBEAT NYSTAGMUS

Acquired downbeat nystagmus is a jerk nystagmus with slow phases that may be linear, accelerating or decelerating (Abel, and colleagues, 1983). As is the case in horizontal nystagmus, downbeat nystagmus may vary in intensity with vertical gaze. This can result in the adoption of a chin-up or chin-down head posture by the patient. By minimizing the nystagmus with such a head posture, oscillopsia is reduced and acuity improved. Prisms (bases up or down) can be used to accomplish the desired vertical gaze angle and increased acuity without the head posture.

Downbeat nystagmus may be induced or enhanced by static tilts in the planes of pitch and roll or the supine position (Baloh and Spooner, 1981; Chambers, and colleagues, 1983; Halmagyi, and colleagues, 1983).

ACQUIRED HEAD OSCILLATIONS

In addition to head oscillations associated with nystagmus, there exist conditions that produce such oscillations in the absence of ocular motor instability. Whereas a non-saccadic oscillation of the eyes is designated nystagmus, a similar oscillation of the head is usually called "tremor." The latter are quite commonly encountered with the most common etiologies being parkinsonism, cerebellar disease, and idiopathic movement disorder designated "benign essential tremor." In the past, many types of head tremor have been called head-nystagmus (Kopfnystagmus) (Klestadt, 1936). When head-nystagmus was a commonly used term, labyrinthine fistula was among the most common etiologies (Stscheglow, 1929) and if the term is to retain any contemporary use, it probably should be limited to the rare head oscillation, accompanying ocular nystagmus, secondary to fistula symptomatology.

The concept of head-nystagmus was said to derive from experiments of pigeons where vestibular stimulation provoked rhythimic movements of the head and only minimal ocular nystagmus (Mygind, 1921). With phylogenetic advancement, the head movements became less and the eye movements predominated. Ontogeny seems to be in parallel since rotational stimulation is much more likely to produce post-rotational head movements in human infants than in children or adults (Mygind, 1921). Nevertheless, pendular movements of the head are observed following both rotation and intense optokinetic stimulation in adults (Schmidt and Schmidt, 1978). Indeed, these authors demonstrated regular electromyographic activity in neck muscles, despite the absence of gross head movement, during routine optokinetic stimulation. This "secondary" nuchal activity associated with ocular nystagmus undoubtedly involves pathways and mechanisms that underlie the spontaneous head oscillations in CN.

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