11. Nystagmus and other ocular motor oscillations

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INTRODUCTION

In Volume 1, this chapter (1) contained a definition and at least one relevant reference for each type of nystagmus and for each type of saccadic oscillation. Table 1 lists the different types of nystagmus along with other terms used in the literature to describe each type; Table 2 does the same for saccadic oscillations. I have developed the definitions and categorizations used herein, initially presented in Volume 1, by the systematic application of criteria derived from accurate ocular motility recordings made during the past 15 years by myself and other investigators. Such criteria clearly differentiate between nystagmus and other ocular motor oscillations and, as a result, some terms containing the word 'nystagmus' were found to be saccadic oscillations. Most oscillations were originally named without the benefits of accurate ocular motor recordings (the naming preceded the understanding). Quotation marks are included to remind the reader that these are *not* truely nystagmus oscillations. Quotation marks are also used for purely subjective clinical terms found in the literature which are inadequate, not clearly defined, or have been misapplied to several different types of oscillations. Since these ambiguous and/or erroneous terms do not convey accurate information about the basic nature of the movement, they are better left at the bedside (preferably under the sheets) and not imposed upon the scientific literature. In those cases where historical precedence supports their continued usage (convergence-retraction 'nystagmus' and voluntary 'nystagmus'), the quotation marks are used to indicate a realization that these are non-nystagmic oscillations (i.e. saccadic). I am hopeful, for the sake of the reader, that future investigators will be more careful about their use of subjective clinical expressions to describe oscillating eyes and will take more care to properly identify an oscillation as one of the main (numbered) types listed in Tables 1 and 2. Obviously, this can only be done with accurate eye-movement recordings as a basis.

Failure to recognize the futility of attempts at clinical descriptions of complex oscillations will result in a continuation of the confusion and contradiction which have permeated the nystagmus literature for the past 100 years. The trained ophthalmological observer is no more able to properly characterize an ocular motor oscillation by visual inspection alone than the trained cardiologist is able to draw a QRS complex without a good instrument to record it. It has been most unfortunate that the eyes are visible to the investigator, for that has been the single largest impediment to good research in this area. Had they been hidden, like the heart, we

TABLE 1. *Nystagmus*

1. Abduction
   'ataxic'
2. Acquired
   fixation
3. Arthrokinetic
   induced
4. Audiokinetic
   induced
5. Bruns
6. Centripetal
7. Cervical
   neck torsion
   vertibral-basilar artery insufficiency
8. Circular / Elliptic / Oblique
   alternating windmill
   circumduction
   diagonal
   elliptic
   gyratory
   oblique
   radiary
9. Congenital
   fixation
   hereditary
10. Convergence
11. Convergence-evoked
12. Dissociated
    disjunctive
13. Downbeat
14. Drug-induced
    barbiturate
    induced
15. Epileptic
16. Flash-induced
    flicker-induced
    induced
17. Gaze-evoked
    gaze-paretic
    deviational
18. Horizontal
19. Induced
20. Intermittent vertical
21. Jerk
22. Latent/Manifest latent
    manifest latent
23. Lateral medullary
24. Lid
25. Miner's*
    occupational
26. Muscle-paretic
    myasthenic
27. Optokinetic
    induced
28. Optokinetic after-
    induced
    postoptokinetic
    reverse postoptokinetic
29. Pendular
30. Periodic/Aperiodic alternating
    alternans
31. Physiological
    end-point
    fatigue
32. Pursuit after-
    induced
33. Pursuit-defect*
34. Rebound
    induced
35. See-saw
36. Spontaneous
37. Torsional
    rotary
38. Uniocular
39. Upbeat
40. Vertical
41. Vestibular
    ageotropic
    alternating current
    caloric
    caloric after-
    compensatory
    electrical
    faradic
    galvanic
    geotropic
    head-shaking
    induced
    labyrinthine
    perverted
    pneumatic/compression
    positional/alcohol
    positioning
    postrotational
    pseudocaloric
    rotational/perrotary
    secondary phase

* may not exist

TABLE 2. *Saccadic oscillations*

1. Bobbing	11. Saccadic double pulses
2. Convergence-retraction 'nystagmus'	saccadic intrusions
'nystagmus' retractoris	12. Saccadic lateropulsion
3. Dynamic overshoot	13. Saccadic pulses
'quiver'	saccadic intrusions
4. Dysmetria	14. Square-wave jerks
5. Flutter	Gegenrücke
6. Flutter dysmetria	hopping 'nystagmus'
7. Macro saccadic oscillations	'lightning eye movements'
8. Macro square-wave jerks	myoclonus
Kippdeviatione/'Kippnystagmus'	saccadic intrusions
'pendular macro-oscillations'	Zickzackbewegungen
saccadic 'nystagmus'	15. Superior oblique myokymia
9. Myoclonus	16. Voluntary 'nystagmus'
'lightning eye movements'	hysterical 'nystagmus'
10. Opsoclonus	'ocular fibrillation'
'dancing eyes'	'ocular shuddering'
'lightning eye movements'	psychological 'nystagmus'
saccadomania	

would be much closer to an understanding of ocular motility than we are at present and perhaps obtaining an OMR (ocular motility recording) would be as familiar to the clinician as an electrocardiogram. Note that I do not use the letters 'ENG' which, in most cases, represent how *not* to record ocular motility, i.e. low band-width, AC-coupled electro-oculogram (EOG) with bitemporal electrodes and curvilinear recording. Such recordings are little better than clinical observation. Good recordings require high band-width, DC-coupling, binocular (*not* bitemporal) electrodes and rectilinear printout. Infrared is more sensitive and more stable than EOG and velocity channels are often an invaluable addition. A blink monitor is also indispensable.

In this volume, only those specific types of oscillation in which noteworthy contributions have been made to the literature in the past 18 months will be discussed. The reader is referred to Volume 1 for definitions and discussions of those types for which no recent contributions to the literature have been made.

NYSTAGMUS

The word 'nystagmus' is derived from the Greek word, *νυσταγμός*, meaning drowsiness, which in turn is derived from *νυστάζειν*, meaning to nod in one's sleep. It should be noted that this nodding oscillation is generated and sustained by the slow downward drifting of the head; the upward head jerks are corrective, i.e. they serve to restore upright head posture.

In keeping with this original definition, nystagmus is defined as follows: a biphasic ocular oscillation containing slow eye movements which are responsible for its genesis and continuation. Fast eye movements (saccades), if they are present, serve a corrective function and do not represent the basic instability. The two phases

of ocular nystagmus are approximately equal in amplitude. This chapter contains discussions of recent studies of 12 nystagmus types from the 41 originally identified in Table 1; they are in alphabetical order.

Circular/elliptic/oblique nystagmus

Circular and elliptic (circumduction, gyratory) nystagmus are forms of pendular nystagmus in which the globe oscillates in a circular path. They should not be confused with torsional (rotary) nystagmus in which the globe itself rotates about an anterior-posterior axis. Rather, they represent the sum of simultaneous horizontal and vertical pendular oscillations which are 90° out of phase. If the amplitudes of the two components are equal, circular nystagmus results; if they are unequal, elliptic nystagmus results. Many times, the nystagmus varies between elliptic and circular and may be dissociated or uniocular. Oblique (diagonal, radiary) nystagmus may be pendular or jerk. If the components are pendular, they are either in phase or 180° out of phase. In oblique jerk nystagmus, the vertical and horizontal components are in phase. These nystagmus types may be congenital or acquired. Acquired, circular/elliptic nystagmus, often dissociated in the two eyes, and almost always associated with truncal or limb ataxia, is usually a manifestation of multiple sclerosis (2). Oblique nystagmus is more commonly acquired than congenital.

A recent paper (3) provides the first good recordings in uniocular elliptical nystagmus. Both horizontal and vertical tracings are shown to document the pendular nature of each component and their 90° phase difference. Velocity tracings also document the pendular nature of the oscillation. Elliptic nystagmus (both binocular and uniocular) has also been associated with internuclear ophthalmoplegia (4). This paper also presents cases of uniocular elliptic nystagmus which are associated with conditions other than multiple sclerosis.

Congenital nystagmus

Congenital nystagmus (CN) is present at birth or shortly thereafter. It may accompany afferent visual defects but is not *caused* by defects in the visual system. Indeed, the oft-quoted association of pendular CN with a sensory defect and jerk CN with a primary motor defect is erroneous. The fact that in many cases one cannot distinguish by clinical observation alone the difference between the several pendular waveforms of CN and the more numerous jerk waveforms further complicates such claims (which were made without eye movement recordings). Systematic ocular motility investigations have revealed no consistent association between waveform and the presence or absence of visual impairment, and both pendular and jerk waveforms have been documented in a single family with hereditary CN (5).

Although the visual deficit is not causal, it can contribute to the intensity of CN. CN is a high gain instability in the slow eye movement subsystem and fixation attempt (the effort to see) is its primary driving force; thus, CN is not a fixation nystagmus. Obviously, poor vision will increase fixation attempts, thereby causing nystagmus intensity to increase. CN can be diagnosed definitely by accurate eye movement recordings. The slow phases of jerk CN are increasing-velocity

exponentials. Three pendular (pendular, asymmetric pendular, and pendular with foveating saccades), 8 jerk (jerk, jerk with extended foveation, pseudo-cycloid, pseudo-jerk, pseudo-pendular, pseudo-pendular with foveating saccades, triangular, and bidirectional jerk), and one combination of pendular and jerk (dual jerk) waveforms have been identified as CN waveforms (6). CN usually damps with convergence and, in many cases, a gaze angle can be found at which the intensity is minimal. Therapeutically, the nulling with convergence may be exploited by the use of base-out prisms and the null angle may be exploited either by prisms or, if the null angle is far removed from primary, by corrective surgery.

The presence of a reversed optokinetic nystagmus (OKN) response or the absence of an OKN response has long been recognized as a pathognomonic sign of CN. Until recently, however, no good explanation for this phenomenon was available. It has also been well known that the intensity of congenital nystagmus varies with gaze angle; indeed, it is this phenomenon which is exploited in prism or surgical therapy. However, a little-known phenomenon is the dependence of null angle upon eye velocity (unpublished observations). Halmagyi et al. (7) documented the latter phenomenon and made the connection to the important clinical observation of reversed OKN. This well-written paper clearly shows how the movement-induced shift in CN null angle is responsible for the appearance of a reversed OKN. The waveform of the reversed OKN is actually the CN waveform that is idiosyncratic for the patient. The importance of recognizing these waveforms is stressed in this paper, since many of the patients studied by this group were not known *a priori* to have CN. Recognition of CN can save patients from many invasive and sometimes dangerous neurological tests. These authors not only made the key observation that attempted smooth pursuit shifts the CN null angle in a direction opposite to the motion of the eyes, but combined it with their study of reversed OKN to reveal this phenomenon as the underlying mechanism for the reversed OKN.

Another recent study (8) of OKN response in patients with CN has come to quite different and, in my opinion, erroneous conclusions. There are 3 serious defects in this paper: one is in the methodology; one is in the method of data analysis; and one is in the inferences drawn in the discussion. The error in methodology stems from the misuse of a very simple computer algorithm which linearizes the slow phase of the nystagmus that is being measured and uses this linearization to obtain a mean slow-phase velocity from which a 'gain' is calculated. The waveforms of a patient with CN are extremely complex and irregular. As the paper discussed above has demonstrated, the response of a CN patient to an OKN stimulus consists of his ongoing CN waveforms. The use of a linearizing algorithm to describe a complex CN waveform is guaranteed to produce erroneous data. While the algorithm in question undoubtedly works well for linear slow-phase waveforms such as normal OKN or normal vestibular nystagmus, it cannot be used to adequately describe such complex waveforms as are found in CN. Thus, the quantities labeled as 'gain', found for CN patients by this method, are virtually meaningless. Furthermore, the great variability of CN waveforms is such that one cannot even hope for a relatively constant error when such linearizations are applied to one waveform and then compared to another CN waveform. Thus, I cannot agree with the authors' assertion that their method of calculating gain is a 'useful approximation of the function of the OKN system'. The application of this or similar types of algorithm

to studies of patients with CN, an application for which they were not designed and are incapable of, is a classic example of the dangers of using a computer inappropriately. My second objection to this paper is the arbitrary and erroneous division of the patients studied into two non-existent groups: sensory-defect and motor-defect. Since all CN is due to a high gain instability in the ocular motor system, *all* such patients have a motor defect. If a subgroup of CN patients happens to have an additional sensory defect, that defect does not change the basic cause of their nystagmus, which is still a motor defect. If a CN patient had a broken arm, one would not call it 'broken-arm nystagmus'. These terms were originally put forth with the specific implication that sensory-defect CN was a nystagmus *caused* by a sensory defect. This has been proved to be erroneous and is recognized to be erroneous by the authors themselves. The fact that 99.9% of ophthalmologists still believe this fairy tale is reason enough to object to the use of these terms; they are neither scientifically nor clinically useful. The study of differences that may exist between patients with CN who have no associated afferent defect and those with an associated afferent defect is not being questioned here; the use of misleading terms is. My final objection to this paper is its main conclusion. Patients with CN either do not respond to OKN stimuli at all or, as we have seen from the previous paper, they respond with an apparent reversal. This reversal was found to be their own idiosyncratic CN waveform beating in a direction dictated by the shift in their null. To adequately stimulate the OKN system, one must provide images moving slowly across the retina in a given direction. For a patient with CN whose slow phase velocity can be several hundred degrees per second, merely placing him in an optokinetic drum does not accomplish this. It is not suprising that most patients with CN do not respond to this stimulus; those who do, are the exception. To adequately stimulate the optokinetic system of a patient with CN, one would have to stabilize the surround on their retinas and then provide that stabilized image with a constant drift velocity. The authors of this paper are aware of this, as indicated by their statements in the methods section. However, they state without evidence that their method of testing the OKN system is adequate. Since they have not, by their own admission, provided the appropriate stimulus to test the optokinetic system of patients with CN, their conclusion that there exists a basic deficit in the OKN systems of patients with CN is unjustified; if such a deficit does exist, it is still to be demonstrated. This paper is not recommended reading but, since it comes from a group who have in the past made worthwhile contributions to the literature, it is important to make potential readers aware of the pitfalls it contains. The authors' confidence in their algorithm has led them to use it indiscriminately without critically evaluating its limitations. There is a strong temptation to trust computer printouts without critically reviewing the algorithms responsible for their generation. This temptation must be resisted by both investigators and those who review manuscripts which make use of such printouts. This paper is an example of a case where an evaluation of the algorithm's capability for generating meaningful data was not performed by either. The algorithm does its job well for those waveforms for which it was designed; it cannot do a good job for most CN waveforms.

A recent paper by Abadi and King-Smith (9) demonstrated that the continuous motion of the eyes in one meridian of patients with CN caused an orientation

amblyopia. To eliminate the effects of eye motion, the stimulus was flashed for 0.2 msec. Three patients with horizontal CN showed a lowered vertical sensitivity and one patient with vertical CN showed a lowered horizontal sensitivity. Another study of the afferent system of patients with CN revealed a marked asymmetry in visually evoked responses (10). In a family with hereditary CN affecting 15 members over at least 5 generations, 2 members were tested: a mother and son. Neither had signs of an abnormality involving the visual pathways. Changes found in visually evoked responses were similar to those found in human albinos who are known to have abnormal retino-striate projections and also nystagmus; neither of the 2 patients tested had evidence of general or ocular albinism. Since the CN waveforms of albinos are indistinguishable from those of non-albinotic CN patients, this study suggests that some patients with CN may also have a related abnormality in their retino-striate projections. This is an area which requires more investigation.

A recent study of CN among the Red-skins of the highlands of Papua New Guinea has revealed that this particular pigmentary abnormality is associated with CN but not with abnormal vision (11). In albinos, nystagmus is invariable and is always associated with poor acuity; among the Red-skins, nystagmus is present in two-thirds and is unrelated to acuity. Although the gene responsible for the inheritance of CN is closely associated with that controlling skin color, it has not been established whether the same or separate genes are involved.

A recent paper presents the results of studies made on a nystagmus patient who, in spite of the history given, can be diagnosed as having CN on the basis of his eye movement waveforms (12). Although the author failed to recognize the nystagmus as being CN, he does claim a number of 'firsts'; the key one is that this study marks the 'first' time that eye movements and acuity have been accurately studied with respect to head position. The author conveniently overlooks studies of such quantities with respect to gaze angle, which is the relevant parameter (13, 14). Other 'discoveries' of previously reported phenomena include the description of 'runaway' nystagmus (known to most as 'jerk with extended foveation') (6) and the variability of CN with effort to see (5). It is unfortunate that the author has taken previously well-documented findings, relabeled them, and presented them as new discoveries. The scientific literature is voluminous enough without filling it with multiple reports of the same phenomena. It is hardly good form to claim as one's own discoveries, results reported 7 years earlier by others. Although the primary responsibility for this paper lies with its author, some responsibility must be shared by the anonymous (somnolent?) reviewers who allowed it to see the light of day; they clearly failed in their responsibility to their Journal and to their colleagues (to us, the readers). The time it takes to read this paper is better spent reading the original works; having to review it and obtaining other opinions were most distasteful.

The first study of the effects of wearing contact lenses on CN has recently been published (15). The contact lenses caused a slight increase in visual acuity when compared to that measured with the patient's previously worn spectacles. More importantly, by measuring the contrast sensitivity function of this patient, it was found that both sensitivity and resolution were improved by the contact lenses. The patient was a myope and, since a myope must accommodate and converge more with contact lenses than with spectacles, the author theorized that the extra vergence effort was responsible for the reduction in nystagmus intensity. This is unlikely,

because the amount of convergence necessary to dampen CN far exceeds the little-added convergence required by contact lenses. Another interesting theory put forward in this paper is that it is possible that information about the presence of the nystagmus reaches the brain via the lid proprioceptors and, thus, it is possible that the nystagmus disturbance itself was modified by the introduction of the contact lens. This is a plausible hypothesis, since one can actually feel the oscillations of the globe when contact lenses are first fitted (personal observation).

Further studies on the use of auditory biofeedback as a means to treat CN have recently been published (16). Continuing his initial studies using biofeedback, Abadi has shown that biofeedback training results in a decrease in the nystagmus intensity and an increase in visual sensitivity. By means of contrast sensitivity functions, this paper shows an increase in sensitivity in both the horizontal and vertical meridians. Improvement was maintained after training had ceased. The authors of this paper do point out that this technique is a specialized solution to particular aspects of CN and does not offer a general solution that is both practical and simple. Studies are now in progress with the same training schedule applied to young children where the visual system is more adaptable. As a final note, the authors suggest that this technique be extended to detecting and correcting other ocular motor problems such as squints and amblyopia.

Downbeat nystagmus

Downbeat is a vertical jerk nystagmus present in primary position with linear upward slow phases and fast phases beating in the downward direction. It is highly suggestive of a disorder of the cranio-cervical junction such as Arnold-Chiari malformations. Contrary to Alexander's Law, it is not maximum in the extreme of downward gaze, but is usually of maximal intensity when the eyes are deviated laterally and slightly below the horizontal. It has also been described in patients with presumed parenchymal cerebellar disease (17). A defect in downward pursuit has been suggested as the cause of this form of pursuit-defect nystagmus (18).

In a recent paper, Baloh and Spooner concluded that downbeat nystagmus is a type of central vestibular nystagmus (19). They studied 17 patients with downbeat nystagmus of various clinical etiologies. All patients had impaired fixation suppression of their vestibulo-ocular reflex (VOR). The data for this paper were gathered and analyzed using the same computer algorithm discussed in the section on congenital nystagmus. In this case, the algorithm was being used properly to analyze nystagmus which had linear slow phases. Thus, the approximations made by the algorithm were realistic and the gains computed were directly related to the actual gains of the subsystems under study, i.e. pursuit, OKN, and VOR. An important finding was the apparent superimposition of the spontaneous nystagmus on attempted pursuit rather than the selective impairment of downward pursuit. This finding is directly related to the hypothesis that downbeat nystagmus is caused by a pursuit defect and supports those arguments that have been proposed questioning the existence of pursuit-defect nystagmus. Five additional papers have recently been published on downbeat nystagmus, but, unfortunately, they all lacked ocular motility recordings. This precludes any discussion of the actual mechanism of the nystagmus since it cannot be determined what the slow-phase wave shapes were.

Two of the 5 deserve comment because of their clinical implications. Costin et al. (20) described 4 cases of downbeat nystagmus in alcoholics and suggest that this association is not well recognized and may form a distinct clinical entity. All 4 patients had oscillopsia and ataxia of gait. In addition, in 3 of the cases, there was evidence of cerebellar cortical atrophy. The striking feature of the downbeat nystagmus in all 4 patients was a marked increase in nystagmus from a fine nystagmus in primary position to a course nystagmus on lateral gaze; modulation of the downbeat nystagmus with vertical gaze was different in all cases. Faria et al. (21) described a case of downbeat nystagmus where this was the salient manifestation of an Arnold-Chiari malformation. All other reported cases of downbeat nystagmus associated with this condition have shown additional neurological symptoms and signs.

Drug-induced nystagmus

A horizontal or horizontal-rotary jerk nystagmus which is gaze-evoked may be induced by the administration of barbiturate (barbiturate nystagmus), tranquilizer, phenothiazine, and anticonvulsant drugs. Vertical nystagmus is often present on upward gaze but only rarely on downward gaze. The nystagmus may be quite dissociated in the two eyes despite the lack of structural disease. Severe intoxication may result in a horizontal, pendular nystagmus at primary position. Careful history-taking and drug-screening blood studies are essential in evaluating patients with nystagmus.

Kairys and Smith (22) have reported on the influence of pilocarpine and atropine on pentobarbital-induced nystagmus in white rabbits. Unfortunately, there are no recordings of the nystagmus; this precluded valid comparisons with known clinical types. Pilocarpine was found to induce nystagmus or increase in intensity of a pre-existing nystagmus; atropine was found to terminate the nystagmus. It is important to know that the frequency of the nystagmus induced was an order of magnitude (i.e. 0.1) less than the nystagmus found clinically in CN or manifest latent nystagmus (MLN). This is stressed because there is a whole section in this paper called 'clinical relevance' which is highly speculative and is not supported by any hard evidence presented in it. Much is made of the fact that pentobarbital, which induced the nystagmus, depresses activity within the brainstem reticular formation. This is correlated with a statement that CN is supposedly inhibited during attention-demanding activities such as writing. Despite the fact that this statement is supported by an old reference, it is well established that CN is intensified by attention-demanding activities (such as the attempt to fixate). The drug-induced nystagmus discussed in this paper seems to operate in an exactly opposite manner to that involved in CN or MLN. That, coupled with the gross differences in frequency and the lack of any waveforms presented in this paper, precludes any meaningful discussion of clinical relevance to these central types of nystagmus. If there is clinical relevance, it is to drug-induced nystagmus which is found in patients who are taking therapeutic doses of various drugs.

Intermittent vertical nystagmus

The occurrence of intermittent attacks of vertical jerk (downbeat or upbeat)

nystagmus is associated with the rare disorder, familial periodic ataxia (23). The episodic symptoms also include vertigo and ataxia. Intermittent vertical nystagmus has been linked to cerebellar or vestibular dysfunction, brainstem disorders caused by multiple sclerosis, and Arnold-Chiari malformation. The occurrence of rotary, vertical, or dissociated nystagmus has been documented in families affected with this disorder. Conversion from primary-position vertical nystagmus to gaze-evoked horizontal jerk nystagmus has also been noted. In many cases, a mild nystagmus and ataxia persisted between the acute attacks.

A very interesting paper on intermittent downbeat nystagmus has recently been published (24). The unique component of the ocular motor abnormality studied was the intermittent nature of the downbeat nystagmus. This was documented (in a patient with Arnold-Chiari malformation) for the first time with accurate ocular motility recordings, as was the rare reduction of the nystagmus after a decompressive procedure. Unfortunately, what may be the most important finding presented in this paper was apparently unrecognized by the authors and was not discussed. This represents, to my knowledge, the first published recording of an acquired nystagmus (albeit in the vertical direction) with slow-phase waveforms which were of increasing velocity. Increasing-velocity slow phases of horizontal oscillations are pathognomonic of CN. There is a possibility that this is an artifact caused by the particular geometry of the infrared system when used in a vertical mode. The waveform for this type of nystagmus should be studied further.

Latent/manifest latent nystagmus

Latent nystagmus (LN) and manifest latent nystagmus (MLN) are nystagmus types elicited by monocular fixation (25). The nystagmus is jerk with the fast phase towards the viewing eye. Although LN/MLN is usually a congenital form of nystagmus, the slow phase is a decreasing-velocity exponential. Thus, it is not true congenital jerk nystagmus which has an increasing-velocity exponential slow phase. MLN occurs in patients with amblyopia or strabismus who, although viewing with both eyes, are fixing monocularly. The direction of MLN in patients with alternating fixation is always in the direction of the fixing eye. Such patients are usually misdiagnosed as having CN, since the nystagmus is present with both eyes open. Accurate eye-movement recordings are the only way to document the nystagmus and diagnose it properly. Unfortunately, most physicians and researchers are unaware of the important clinical and functional differences between MLN and CN and cannot distinguish between these two separate congenital forms of nystagmus.

Although MLN does not have a null angle (this is shown in Fig. 9 of Ref. 25), the variation of MLN with gaze angle in accordance with Alexander's Law may result in a head turn. Usually the head turn is such that the viewing eye is in adduction. If the patient is an alternate fixer, he may adopt one head turn when viewing with the right eye and an opposite head turn when viewing with the left eye, so that in each condition the viewing eye is in adduction. It becomes very important, therefore, to distinguish MLN from CN before any consideration of surgical intervention is made. Surgery for CN will depend on the location of a true null, whereas surgery for MLN should be performed only if the patient consistently fixes with one eye and accompanies this fixation with a head turn. Alternate fixers who adopt alternate

head turns cannot be helped by surgical rotation. As pointed out in this paper, the situation is complicated by the fact that some patients have neither a pure CN nor a pure MLN; various combinations of the two exist and the only way to accurately diagnose the condition is by means of ocular motility recording and waveform analysis.

Abadi (26) has recently studied pattern contrast thresholds in LN. He found that the shape of the contrast sensitivity functions (CSF) for the nystagmus patients showed no low-frequency attenuation. This was attributed to the induced temporal movements of the retinal image. Since data for only one subject were shown and the eye movement recordings were made with EOG, it is impossible to tell whether any or all of these subjects had MLN. More sensitive methods are sometimes needed to detect nystagmus present under binocular viewing conditions. It would also have been interesting to know whether any of the patients had strabismus, since all of the patients we have studied with MLN had this disorder. In attempting to discuss the possible initiating factors involved in LN, the author arrives at a concept of a switching mechanism which is very similar to the one we have previously suggested (25). It is not clear from the author's explanation how the signal from a binocular comparator associated with deciding which of the two eyes should take up fixation is translated into the ocular motor control signal which causes both eyes to conjugately drift in a given direction. There is little evidence for the existence of two monocular control systems for version eye movements. Anatomical and physiological evidence supports directional control systems which move the eyes conjugately. Our hypothesis of a basic central abnormality involving differences between monocular and binocular ego-direction is supported by this study. This very same comparator, which must utilize monocular visual inputs to decide upon a binocular zero direction (i.e. ego-direction), seems to be where the defect lies.

Optokinetic nystagmus

Optokinetic nystagmus (OKN) is a form of induced jerk nystagmus which is extremely valuable diagnostically. The nystagmus is induced by presenting to the subject a visual pattern which moves with constant velocity in a given direction. The induced eye movements consist of constant-velocity (linear), conjugate eye movements in the direction of the moving stimulus interspaced with fast phases in the opposite direction. OKN testing can be used to document the existence of vision in infants or patients with functional visual loss, to localize cerebral hemispheric lesions, to induce convergence-retraction 'nystagmus', to demonstrate the adduction insufficiency in internuclear ophthalmoparesis, to diagnose ocular motor nerve misdirection, to diagnose ocular myasthenia gravis (by injecting anticholinesterase during the OKN test and noting the velocity of the fast phases) and to diagnose congenital nystagmus (by getting inversion).

Despite the clinical importance of OKN testing, the literature on the subject is as confusing as it is voluminous. There is disagreement among those currently doing OKN research about both the nature of an adequate OKN stimulus and the characteristics of a true OKN response. According to one school of thought, OKN is divided into two types – Stier and Schau nystagmus. This is based on the work of Ter Braak (27) who equated the Stier (field) OKN with subcortical mechanisms,

while the Schau (object) OKN was related to cortical mechanisms. In humans, this is equivalent to full-field versus central-field (foveal) OKN. Clinically, it is the foveal OKN which is stimulated by the familiar OKN tape. The work of Dichgans (28) seems to support the hypothesis that one can simulate a full-field stimulus with a 90° horizontal strip. At the other end of the spectrum, Robinson (29) is of the opinion that true OKN must induce circular vection and be followed by optokinetic after-nystagmus (OKAN). Any stimulus which does not produce OKAN and circular vection is, therefore, inducing a 'pseudo-OKN' which is probably mediated by the pursuit mechanism in humans. Because one cannot dissociate the function of pursuit in the full-field stimulus condition, Robinson proposed that the OKN mechanism can be isolated only by studying OKAN. The picture is further complicated by the interrelation between the vestibular system and the OKN system (30). A paper by Muratore and Zee (31) on pursuit after-nystagmus has raised serious questions about Robinson's definition of OKN. It was found that simple pursuit induced an after-nystagmus. Thus, we are faced with the fact that both full-field stimulation and a simple foveal target, which is pursued, can induce an after-nystagmus and it may be impossible to separate true OKN from pursuit OKN if, indeed, they result from different mechanisms. While it has been presumed that pursuit is a foveal reflex, evidence has been presented (32) that pursuit need not be foveal. It is within the context of these conflicting views on the nature of an adequate stimulus for OKN that one must read the literature on this subject. It is quite possible that, when all the evidence is in, we may find that, despite differing functions and phylogenetic origin, both the optokinetic and the pursuit response are mediated by the same neurophysiological efferent mechanisms and differ only in afferent magnitudes. That is, the magnitude of the following response will be related to the amount of retinal area stimulated and the percentage of that area that is in the direction of motion. Similarly, the ever-present interaction with the vestibular system would be proportional to the following response elicited by the particular stimulus presented. Thus, a small stimulus in the periphery would elicit a weaker response than a stimulus whose image was a visual strip across the retina including the fovea where the long direction of the strip corresponds to image motion. As the stimulus becomes more compelling and the resultant following response harder to suppress, one would move from the 'pursuit domain' to the 'optokinetic domain' without necessarily changing the mechanism responsible for the respective responses. Of course, the retinal sensitivity map will vary from species to species depending upon the morphology of the individual retinas.

In a recent paper (33), it was reported that the combination of a behavioral and a surgical manipulation resulted in the abolition of OKN in the cat. Neither visual deprivation nor surgical section of the optic chiasm (the two manipulations employed) result in abolition of OKN. The combination of these two manipulations however, severs all connections to the nucleus of the optic tract and thus implicated this structure in the generation of OKN. In another study on cats (34), the response of vestibular nucleus neurons to rotation of a large-field visual pattern was studied. These nuclei responded in the absence of the cerebellum, or with lesions in the central tegmental tracts, medial longitudinal fascicles, superior colliculus, or inferior olive. However, bilateral pretectal lesions severely affected the horizontal OKN responses of these neurons and the nystagmus itself. The term 'pretectal'

implies the nucleus of the optic tract and other pretectal nuclei. A major OKN pretectofugal pathway reaches the vestibular nuclei through the area of the nucleus reticularis tegmenti pontis.

Recently Wolfe et al. (35) studied the binocular contribution to OKN in normal and stereoblind subjects. They found that OKN can utilize purely binocular or 'cyclopean' input. The binocularity of this mechanism was not disrupted in subjects who lacked stereopsis. They concluded that more than one binocular process exists in the visual system. There are two recent papers on directional asymmetry of OKN whose significance I find impossible to evaluate. The first paper studied patients with amblyopia (36). No sensory anomalies were found to be the basis for the abnormal OKN responses measured. However, the claims of directional asymmetry in OKN response cannot be evaluated without more knowledge of the presence and type of spontaneous nystagmus that may have been present in these patients. No tracings were shown with the subjects simply fixating, so that it could not be determined whether they had CN or MLN. Without this knowledge (considering the probability that many of these patients had either of these conditions), one cannot evaluate the presence or absence of an OKN response or indeed the appearance of asymmetrical response. CN is not mentioned in the paper, nor is MLN; there is a discussion of LN but, unfortunately, it contains an error. Contrary to what is stated in this paper, and in the reference listed, the slow phase of latent nystagmus is in the direction of the covered eye and is strongest when gaze is directed to the side of the *uncovered* eye. The second paper is on optokinetic asymmetry in patients with maldeveloped foveas (37). Since all of these patients had congenital achromatopsia and CN, evaluation of their OKN responses by a digital computer program incapable of analyzing CN waveforms (using input from bitemporal EOG electrodes) is not justified (see discussion of this problem in the section on Congenital Nystagmus). I am therefore unable to assess the significance of the findings of this paper. A related finding in pigeons was that lesions of the foveal and parifoveal area did *not* impair OKN (38).

In a recent paper, Baloh et al. (39) studied OKN asymmetry in patients with parietal lobe lesions. They found that ipsilateral foveal pursuit was impaired to a greater extent than full-field pursuit. They inferred from this the existence of two separate pathways by which visual signals for OKN slow phases reach the ocular motor centers in the brainstem. Their conclusion was that the ipsilateral OKN deficit seen in their patients resulted from damage to the foveal-pursuit pathway. A method has recently been presented which may increase the sensitivity of OKN testing to various pathological conditions (40). It involves presenting an OKN stimulus at a low speed followed by a step change in velocity to a higher speed and terminating with a step change back down to the initial speed. It is suggested in this paper that these sudden supraoptimal accelerations of the stimulus intensify the OKN responses and that the reactions are varied with different pathological conditions. This is an area which requires more study before it can be properly evaluated. Abel et al. (41) have pointed out that the OKN characteristics depend upon the recording techniques employed. They found that wearing infrared spectacles produced no statistically significant change for 'follow' OKN but did cause a significant decrement in 'stare' OKN. Thus, one must balance the increased accuracy offered by infrared against the possible decrement in response it may cause.

Optokinetic after-nystagmus

Optokinetic after-nystagmus (OKAN) is a continuation of optokinetic nystagmus (OKN) induced after the cessation of visual stimulation in complete darkness; this is known as OKAN I or post-OKN. After variable periods of time, it is followed by OKAN II; this is a secondary OKAN or reverse post-OKN. OKAN I has the same direction as the preceding OKN, whereas OKAN II is in the opposite direction. The duration of OKAN I is variable. In an effort to identify the mechanisms of OKAN I and OKAN II, Waespe et al. (42) studied the effects on OKAN II of brief periods of visual fixation during OKAN I. They found that OKAN I and OKAN II were influenced in a reciprocal way, i.e. OKAN I is reduced when OKAN II is increased. In those human subjects who exhibited no OKAN II in the controlled experiments, the suppression of OKAN I resulted in an OKAN II. They concluded that OKAN II depended upon the parameters of the preceding OKN stimulation and not upon the occurrence of OKAN I. Attempts to inhibit OKAN II by brief fixation periods resulted in a return of the OKAN II and a second maximum in its intensity; OKAN I showed little recovery. They concluded that OKAN II was a sign of central activity or counter-regulation which played a decisive role during all phases of OKAN.

A very interesting, and potentially far-reaching, observation has been recently presented by Collewijn et al. (43). They found that the decay of the slow phase velocity of both OKAN and postrotary nystagmus was best fitted by *linear* and not exponential functions. It can be directly inferred from this that the optokinetic and vestibulo-ocular systems are non-linear. A velocity storage system with a constant discharge rate was postulated as the main non-linear element. In attempting to compare the discharge rate to the exponential curves, apparent time constants were defined as the time taken to decay to 37% of initial velocity. While the utility of such a time constant is debatable (I prefer using the slope of the straight line), it in no way detracts from the very important observations presented in this paper. The authors cite numerous studies done on monkey to show that this is not limited to the rabbit OKN system. In another paper, Skavenski et al. (44) studied OKN and postrotary nystagmus before and after 20 habituating exposures to either vestibular or optokinetic stimulation. Their aim was to test whether both reflexes shared the same velocity storage mechanism. It is interesting in the context of the previous paper to note that they fitted the time courses for the decay of eye speed with straight lines.

Periodic/aperiodic alternating nystagmus

Periodic alternating nystagmus (PAN) (nystagmus alternans) is an extraordinary phenomenon in which a persisting horizontal jerk nystagmus periodically changes direction. There may be a fixed sequence consisting of approximately 90 sec of nystagmus beating in one direction, 10 sec of neutral phase in which the eyes stop, beat downward irregularly, or oscillate pendularly, followed by 90 sec of beating in the opposite direction. In many patients, the timing is very asymmetric, but since the reversals continue to occur, it may be considered aperiodic alternating nystagmus (APAN). The waveforms of the slow phases will depend on the etiology in each case. PAN can be conceptualized as resulting from periodic shifts of the null zone of a manifest horizontal jerk nystagmus (45). PAN/APAN have been associated with

congenital nystagmus, head trauma, encephalitis, syphilis, multiple sclerosis, spinocerebellar degenerations, and posterior fossa tumors and infarction.

A recent paper on PAN revealed that during the nystagmus-free intervals (neutral phase), refixation saccades and pursuit movements were abnormal (46). Saccades were hypermetric and pursuit movements were saccadic with occasional hypermetric saccades interspersed. The movements were distinctly cerebellar in character and these findings support the hypothesis that PAN is due to hyperexcitability of the vestibular nuclei on either side of the brainstem caused by loss of cerebellar inhibition. In another recent paper on PAN, Halmagyi et al. (47) report successful treatment using the drug baclofen. This drug abolished the nystagmus and relieved the oscillopsia in 2 patients with acquired PAN, but it was ineffective in another with congenital PAN. Although the mechanism of action of baclofen on PAN is unknown at present, the discovery of its utility is significant.

Pursuit-defect nystagmus

Pursuit-defect nystagmus may be a vertical or horizontal jerk nystagmus and is supposedly caused by a unilateral defect in pursuit. Thus, a defect in downward pursuit would result in a drifting of the eye upward and give rise to downbeat nystagmus. Similarly, a defect in upward pursuit would cause upbeat nystagmus and a defect in pursuit to the left or right would cause a left-beating or right-beating horizontal pursuit-defect nystagmus (48). The slow phases of pursuit-defect nystagmus are linear. In a recent letter (49), we have questioned the whole concept of 'pursuit-defect' nystagmus. To document the absence of pursuit in the presence of an ongoing nystagmus, we feel it is insufficient to show that the ongoing slow-phase direction is not reversed by pursuit in the other direction. For pursuit to be regarded as totally absent, no change in the slope of the slow phase should result when pursuit is attempted in the other direction.

A recent paper on downbeat nystagmus by Baloh and Spooner (19) supports our contention that the pursuit asymmetry seen with spontaneous nystagmus may be the result of the nystagmus rather than its cause. They concluded that downbeat nystagmus was a type of central vestibular nystagmus rather than a pursuit-defect nystagmus. Of the 3 patients shown in their Figure 3, one was unable to change the upward slow phase upon attempted downward pursuit, another was able to modulate the upward slow phase, and a third was actually able to reverse the upward slow phase and produce downward smooth movements. Thus, we have 3 patients afflicted with downbeat nystagmus and only 1 patient was unable to modify the upward slow phases with attempted downward pursuit. The fact that 2 of the patients could alter the upward slow phases of their ongoing nystagmus by attempted downward pursuit clearly argues against the hypothesis that it is the absence of downward pursuit that caused the nystagmus. Both our observations and the findings of Baloh and Spooner argue effectively against the existence of pursuit-defect nystagmus. The original assumption of an absence of pursuit or the recent hypothesis of a directional preponderance of the pursuit system (50) do not stand up to these recent observations that have been made during pursuit attempts by patients with ongoing nystagmus.

Rebound nystagmus

Rebound nystagmus is a gaze-evoked horizontal jerk nystagmus which fatigues and changes direction with sustained lateral gaze and/or horizontal gaze-evoked nystagmus which, upon refixation to primary position, transiently beats in the opposite direction (51). The slow phases are decreasing-velocity exponentials. Rebound nystagmus is often present in patients with parenchymal cerebellar disease, but normal subjects may demonstrate rebound nystagmus after prolonged far lateral gaze if the lights are shut off the moment the eyes are returned to primary position. Rebound nystagmus may be mistaken for periodic alternating nystagmus with asymmetric cycles (APAN).

Yamazaki and Zee (52) have recently studied rebound nystagmus in a patient with a floccular tumor. When the tumor was confined to the cerebellar flocculus, the ocular motor signs included impaired smooth pursuit, impaired cancellation of the vestibulo-ocular reflex, and gaze-paretic and rebound nystagmus. When the tumor invaded the brainstem in the region near the vestibular nuclei however, the rebound nystagmus disappeared; the other ocular motor abnormalities remained. The authors concluded that the floccular lesions unmasked a bias which created rebound nystagmus and that the bias probably arose in the vestibular nuclei. Upon studying the patient 20 days after an operation to remove the tumor, the patient was found to have Bruns nystagmus (1). Analyzing their findings, the authors concluded that the bias that creates rebound nystagmus is developed in the brainstem and not in the flocculus. The exact mechanisms involved in creating this bias are still not understood.

Vestibular nystagmus

Vestibular (labyrinthine) nystagmus is a jerk nystagmus which may be acquired due to central vestibular dysfunction, a peripheral (end-organ) vestibular disease or vestibular system plasticity reacting to dysfunction and producing compensatory nystagmus. It also may be induced (alternating current, caloric after, electrical, faradic, galvanic, perverted, pneumatic/compressive, positional/alcohol, post-rotational, pseudo-caloric, or rotational/perrotary). Pathological vestibular nystagmus may be spontaneous or may be induced by having the patient adopt certain positions (positional) or shaking his head (head shaking); in some patients, the act of changing positions induces the nystagmus rather than the position finally achieved (positioning). Positional nystagmus may beat constantly in the direction of the ground (geotropic) or it may beat in the direction opposite to the ground (ageotropic) regardless of head position. The slow phase of primary-position vestibular nystagmus is linear and the nystagmus increases with gaze towards the fast phase in accordance with Alexander's Law. Vertigo usually coexists with the nystagmus. Acute lesions of the cerebellar flocculus (the vestibular cerebellum) can produce a similar nystagmus. In normal subjects, some degree of vestibular nystagmus may be induced when the labyrinth is stimulated with warm or cold water applied to the tympanic membrane. Direction of this nystagmus is such that the fast phase beats opposite to the side in which cold water is applied or in the same direction as the side in which warm water is applied. Caloric nystagmus and caloric

after-nystagmus (also called secondary-phase nystagmus) coexist with vertigo and past pointing. The direction of the vertiginous environmental movement (circular vection) is in the direction of the fast phase of the nystagmus. Pseudo-caloric nystagmus is an appropriate cold caloric and an inappropriate warm caloric response from an ear with abolished vestibular function. Vestibular nystagmus is associated with Ménière's disease and many disease processes of vestibular end-organ or nerve. Spontaneous vestibular nystagmus is directed to the side opposite the lesion.

Two recent papers discussed the benefits and drawbacks of air versus water caloric irrigations. Nijhuis and Huygen (53) found that the variability of single responses was correlated with the level of the response. Allowing for that correlation, they found no difference in variability between the two types of caloric stimulation. Zangemeister and Bock (54) point out that air caloric irrigation is more convenient for both patient and operator. They stressed the need for a high flow rate and reproducible tip position when performing air irrigation and also stated that the actual temperature should be measured at the irrigation tip. They found a higher standard deviation for air irrigations, but this may be due to the fact that they did not correlate standard deviation with magnitude of the response. While it seems clear from both these papers that good results can be obtained with both air and water, it is apparent that the techniques must be carefully controlled when using air. The effects of 6 different conditions of fixation on caloric nystagmus were studied by Karlsen et al. (55). They found that caloric testing is best carried out in darkness or with the eyes closed; specifically, the use of +20-diopter lenses should be avoided.

Boumans et al. (56) have recently published the results of a study on the effects of step changes in angular velocity on the resulting nystagmus. They calculated a time constant from the slope of the decay of slow-phase velocity after a step change in angular velocity. Their results showed: a test-retest variability which was greater than the inter-subject variability; significant differences in mean values of time constants for clockwise and counterclockwise rotation; no relation between the time constant and the amplitude of the step of angular velocity; and a linear increase for the maximum slow-phase eye velocity with amplitude for velocities below 100° per second, whereas at higher velocities, saturation occurred. These findings imply linearity in the system and must be reconciled with those previously mentioned by Collewijn et al. (43). Gorman and Pyfer (57) have studied the post-rotatory nystagmus responses of adults, normal children, learning disabled children and emotionally handicapped children. They found that the 3 child populations had characteristically different nystagmus responses from that of the adult group and that the learning disabled and the emotionally handicapped child populations had significantly different responses from those of the normal child population. However, the two handicapped populations did not significantly differ. These authors, although using primitive ocular motility recording equipment, did take care to maintain low light levels and instructed their subjects to count backwards following the cessation of rotation. There seems to be great interest in studying the vestibular responses of handicapped children, but, unfortunately, many of those involved in this endeavor are not carefully recording the eye movements (they are relying on visual observation) or are not maintaining the proper conditions for good vestibular testing. Igarashi et al. (58)

studied the effects of off-vertical tilt and macular ablation on postrotatory nystagmus in the squirrel monkey. They found a significant difference in the nystagmus decay curve by tilting the rotation axis and, by means of macular ablation, established that it was these gravity receptors which were responsible for the differences.

Kato et al. (59) have recently studied visual suppression of caloric nystagmus in cats. In monkeys with lesions of the flocculus, loss of visual suppression is accompanied by disturbance of OKN. Lesions of the cat inferior olive did not cause loss of visual suppression of caloric nystagmus or interference with OKN. However, lesions of the superior colliculus resulted in a loss of visual suppression and an interference in OKN response. These authors conclude that the superior colliculus may be an important relay nucleus to the flocculus in conveying visual signals responsible for the VOR gain. Buettner and Büttner (60) studied vestibular nuclei activity in monkey during suppression of both vestibular and optokinetic nystagmus. Suppression of OKN attenuated the neural activity in this area and suppression of vestibular nystagmus shortened the time constants of the decay of the neuronal activity. The authors postulated a feedback circuit which normally operates during both types of nystagmus but which is interrupted by suppression. It should be pointed out that this is a linear model which must be reconciled with the recent findings of Collewijn et al. (43). Blair and Gavin (61) studied the effects of sagittal incision of the macaque brainstem near the midline and caudal and ventral to the abducens nuclei. They found that this induced a syndrome which included reduction of the time constant of vestibular nystagmus and reduction in OKAN. Their results suggested that inhibitory commissural fibers in the caudal brainstem influenced neural circuits that normally transform the short cupular time constant into the long nystagmus time constant. They suggest that the neuronal linkage involved in this transformation is under both excitatory (from the cerebellar vermis via the fastigial nuclei) and inhibitory (vestibular commissural fibers) control. If the latter fibers are important in the control of the vestibular time constant, then a great variety of lesions in the brainstem will result in reductions in this time constant to values near that of the cupular time constant; this important clinical point is made in the paper.

There have been several recent papers worth mentioning because of the clinical information they contain. Dohlman (62) studied the mechanism producing the symptoms of Ménière's disease. Characteristic symptoms of this disease include nystagmus beating in an ipsilateral direction eventually changing to a contralateral direction; normal function returns between attacks. Since all of these symptoms have been reproduced in animals by increasing the potassium concentration of the interstitial fluids surrounding the afferent nerve branches from the sensory areas of the inner ear, a mechanistic explanation is offered based on this finding. A low potassium concentration, which increases the action potential frequency, would explain the appearance of an ipsilateral nystagmus. Raising the concentration further depresses the action potential frequency and results in a contralateral nystagmus. Reker (63) recently commented on the reproducibility of two methodologies employed in studying peripheral vestibular nystagmus. Although the maximum intensity of a spontaneous nystagmus is several times higher with eyes closed and under Frenzel's glasses, and a weak nystagmus is difficult to recognize

with the latter, the spontaneous nystagmus due to purely peripheral lesions shows large intensity fluctuations with closed eyes. Contrary to this, the use of Frenzel's lenses results in very little fluctuation. Thus, although the absolute reaction is lower, the content of information is higher when using Frenzel's glasses because of the better reproducibility and this method is recommended for use with patients. Minnigerode and Meissner (64) have recently reported on returning function after sudden unilateral isolated vestibular loss. The interesting observation they make is that function may return with or without recovery nystagmus with a numerically nearly equal frequency. Finally, Brandt and Daroff (65) recently wrote an excellent review article on multisensory physiological and pathological vertigo syndromes. In this paper, not only are mechanisms for the various types of vertigo discussed but also several types of nystagmus are mentioned with other clinical signs. This paper is recommended reading. Also recommended reading in the area of vestibular and optokinetic nystagmus and their interaction is the chapter on nystagmus and pursuit eye movements in the recent *Neurosciences Research Progress Bulletin* by Henn et al. (66).

A recent study on the maturation of vestibular nystagmus in infancy and childhood is important in studies of normals as well as patients (i.e. children with learning disabilities) (67). Ornitz and co-workers found significant changes in nystagmus parameters in respect to maturation. Young infants have larger-amplitude, higher-velocity beats than older children during both primary and secondary nystagmus. The parameters describing both types of nystagmus reached their peak values and terminated earlier in the infant than in the older child. The secondary-nystagmus/primary-nystagmus ratio was significantly greater in early infancy. These factors should be taken into account in any studies of children whether they be normal or patients. Although it is well known that vigilance is important when measuring nystagmus, Kileny et al. (68) have recently found that conversation was consistently superior to mental arithmetic when measuring caloric nystagmus. Since mental arithmetic is the method employed by most laboratories, it would be interesting to investigate the relative merit of conversation when measuring various types of vestibular nystagmus. Finally, there have been 3 recent papers on the use of computers in automatic analysis of nystagmus (69–71). Because these programs are designed for specific applications, I will not discuss the details here. However, they all use linearizing algorithms for the slow phases and the same warning previously mentioned with regard to several papers reviewed in this chapter must be reiterated. These programs will *not* work for CN waveforms. They will work well for normal vestibular nystagmus and for OKN.

OTHER OCULAR MOTOR OSCILLATIONS

Non-nystagmic ocular motor oscillations represent solely saccadic or saccadically initiated instabilities. I have identified 16 varieties of saccadic oscillations which have been characterized in the literature by 32 different terms, including 8 which erroneously contain the term 'nystagmus'. This chapter contains discussions of recent studies of 3 types of saccadic oscillations from the 16 originally identified in Table 2; they are in alphabetical order.

Opsoclonus

Opsoclonus (saccadomania) consists of rapid, involuntary, chaotic, repetitive, unpredictable, conjugate saccadic eye movements in all directions which prevent fixation and persist during sleep. The terms 'dancing eyes' and 'lightning eye movements' have been used to describe the eye movements of patients with opsoclonus.

Recently, Gresty et al. (72) studied the mechanism of certain 'rotatory' eye movements in opsoclonus. Although the paper has some interesting findings, there are errors in terminology. The eye movements which are presented in the paper are actually elliptical rather than rotatory. Rotary (rotatory) eye movements are synonymous with torsional eye movements. What the paper deals with, therefore, are bursts of elliptical saccadic eye movements. The error in terminology was the introduction of the description 'rotatory nystagmus' for these eye movements; the movements are saccadic and, therefore, not nystagmus. Torsional (rotary) nystagmus consists of torsional slow movements of the globes about their anteroposterior axis; they are continuous and do not occur in sporadic bursts. Similarly, elliptic nystagmus is a continuous rotation of the eyes in an elliptical pattern. Thus, references to rotatory nystagmus in this paper should be disregarded and the reader's attention should be directed to the movements themselves and the explanation offered. Basically, what has been shown is that opsoclonus may consist not only of horizontal, vertical, and oblique saccades, but also of back-to-back curved saccades. The latter consist of bursts of horizontal and vertical flutter with small phase differences. This is indeed an interesting finding and it is unfortunate that the paper confounded the term 'rotatory nystagmus'. Such nystagmus is slow eye movement induced and should not be classified with flutter and opsoclonus as suggested in this paper.

Square-wave jerks

Square-wave jerks (SWJ) (Gegenrücke, hopping 'nystagmus', 'lightning eye movements', and Zickzackbewegungen) consist of a pair of saccades which initially take the eyes off fixation by a few degrees and after a suitable latency (about 200 msec) return the eyes to the target. SWJ intrusions may occur in normals (especially upon closure of the eyelids) or may represent pathology suggestive of cerebellar disease.

Recently, Herishanu and Sharpe (73) studied the occurrence of SWJs in the normal population. They found that their frequency in the elderly was significantly higher than in young subjects. Their results suggested that SWJs more frequent than 9/min in young patients can be considered abnormal. This figure is important clinically, since it has long been recognized that SWJs occur both in normals and in patients and that no clear dividing line has been available. Frequent SWJs are usually seen in patients with other cerebellar signs. Perhaps the occurrence of frequent SWJs precedes the more common and more debilitating signs that are found in cerebellar system disease.

Voluntary 'nystagmus'

Voluntary (hysterical, psychological) 'nystagmus' is not nystagmus at all but a series

of back-to-back saccades, interrupting fixation, whose timing is such that the waveform traced out appears to be pendular (i.e. a voluntary flutter) (74). The frequency of this oscillation, also called 'ocular fibrillation' and 'ocular shuddering', is typically 8–23 Hz. The oscillation is horizontal, conjugate, and each burst usually has a duration of less than 30 sec.

Nagle et al. (75) have studied saccadic suppression associated with voluntary 'nystagmus'. They found that saccadic suppression was similar during voluntary 'nystagmus' and during voluntary saccades, as would be expected given the saccadic nature of this oscillation. However, the failure of their subjects to perceive stabilization of the visual world during voluntary 'nystagmus' differs from the stabilization that is perceived during a voluntary saccade. Since the subjects also failed to perceive movement of a stabilized retinal image during voluntary 'nystagmus', no extraretinal signal was influencing the apparent stimulus position. This result contradicts the hypothesized role of saccadic suppression and subjective stabilization. The suppression finding points out a significant similarity between the two types of eye movement, while the difference in subjective stabilization reveals a significant distinction between them. Nagle and co-workers concluded that saccadic suppression is not adequate by itself to prevent the subjective appearance of movement in the visual world during saccades and that it is related to the mechanism of saccadic generation itself. Stabilization was linked by these authors to the act of re-targeting rather than the generation of a saccade.

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