Strabismus — a necessary condition for latent and manifest latent nystagmus*

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ABSTRACT. Our previous study of 31 cases of manifest latent nystagmus (MLN) suggested a one-to-one relationship between the occurrence of MLN and the presence of strabismus. This study of three patients with both latent nystagmus (LN) and MLN has elucidated the relationship between strabismus and these two intimately related types of nystagmus. Both LN and MLN have jerk waveforms whose fast phases are in the direction of the fixating eye and whose slow phases are decreasing-velocity exponentials; the nystagmus is simultaneously present in the two eyes. LN and MLN should not be confused with congenital nystagmus which differs in waveform, variability with gaze angle, mechanism, clinical signs and therapy. During binocular viewing, if the two eyes are aligned with normal retinal correspondence, the patient with true LN will have no nystagmus. Only if one of his eyes assumes a tropic position, might MLN develop. Under cover, the development of a phoria is synchronous with the occurrence of LN. Thus, strabismus in the form of a tropia or a phoria emerges as a necessary, but not sufficient, condition for both LN and MLN. That is, during the time when a nystagmus is present, the non-fixating eye (the eye behind cover or which is being suppressed) will be phoric or tropic, respectively. In LN cases with abnormal retinal correspondence, although no nystagmus may be present with the eyes in their normal fixed tropic position, cover will induce nystagmus along with a phoria. The intriguing and highly desirable possibility of converting MLN to LN, in a very young child with a fixed tropia, by early strabismus surgery is discussed.

Key words: strabismus; latent nystagmus, manifest latent nystagmus; nystagmus

INTRODUCTION

True latent nystagmus (LN) is a congenital type of jerk nystagmus present upon covering, blurring, or reducing the image brightness in one eye and absent with both eyes fixating. The direction of the fast phase is always towards the viewing eye. The slow phases have a decreasing-velocity exponential shape. This type of slow phase is not present in other types of primary position horizontal nystagmus (Dell’Osso et al. Neuro-ophthalmology 1983, Vol. 3, No. 4, pp. 247-257)
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and is different from the jerk congenital nystagmus (CN) waveforms which have increasing-velocity slow phases (Dell'Oso & Daroff, 1975). LN, while congenital, should not be confused with nor mis-identified as CN; they are different in mechanism, waveform, variation with gaze angle, clinical signs and therapy.

Strabismus is defined as a squint or lack of parallelism of the visual axes of the eyes. Our use of the term includes both tropias and phorias and does not differentiate between sub-groups of strabismus patients with differing sensory abnormalities. The ocular motor observations presented herein relate ocular alignment to the presence or absence of nystagmus and are, in that sense, independent of specific sensory defects. In subjects with strabismus who, although viewing with both eyes (i.e., both eyes are open), fixate monocularly, there is cortical suppression, an analogous situation to occlusion. In these cases, the ongoing nystagmus (if present and not CN) is actually manifest latent nystagmus (MLN) (Dell'Oso et al., 1979). MLN is a jerk nystagmus with fast phases towards the fixating eye and slow phases that are decreasing-velocity exponentials; MLN is identical to LN in waveform and mechanism and, as the name implies, differs only in its manifestation with both eyes open. A diagnosis of pure (or true) LN is used when a patient exhibits no nystagmus with both eyes open and covering of either causes LN to develop. Patients with MLN also exhibit the proper direction of LN upon covering of either eye. Finally, when both eyes are open, a patient may have no nystagmus at times (i.e., true LN) and manifest nystagmus at other times (i.e., MLN). It is such patients who are presented here.

Until now, there have not been any published reports of true LN which documented its existence (i.e., no nystagmus with both eyes viewing and jerk nystagmus in the direction of the viewing eye with other eye covered) by showing simultaneous binocular eye movement recordings made with quantitative, sensitive, low-noise, drift-free recording techniques. Using such recordings, we found the presence of small-amplitude MLN in patients who did not appear to have nystagmus during binocular viewing and were, therefore, clinically diagnosed as having LN (Dell'Oso et al., 1979). Since LN usually is a clinical diagnosis made without good oculographic records, our study of MLN caused us to question the existence of true LN; we had never seen it in the records of over 300 cases of congenital varieties of nystagmus. To the best of our knowledge the first, and only other, recording of a patient with LN that clearly shows the absence of nystagmus under binocular viewing conditions is contained in the doctoral dissertation of K. J. Ciuflreda (1977); it was the first such recording using accurate equipment. Unfortunately, since only the viewing eye was shown under conditions of monocular fixation, no data are available about the position of the covered eye.

We report here three cases, all of whom were documented by quantitative eye movement recordings. Each had both pure latent nystagmus (that is, no nystagmus present when both eyes were open and aligned on the target) and MLN (which appeared with both eyes open when one eye became mis-aligned). Our recordings confirm the long-suspected causal link between strabismus and the occurrence of LN and MLN, and establish the existence of true
LN as a distinct, albeit rare, entity. These three LN patients are the only ones we have found in the past decade of recording over 350 patients referred to us with congenital types of nystagmus; in contrast, 31 were shown to have MLN (Dell’Osso et al., 1979). We never have recorded a patient who exhibited true LN at all times.

METHODS

Horizontal eye movement recordings were made using infrared oculography with a full-system bandwidth (position and velocity) of DC to 100 Hz (Biometrics Model-200 and a rectilinear Beckman Type-R Dynograph; both modified to achieve the above bandwidths). Sensitivity was better than 0.5 deg, noise less than 0.25 deg p-p, and drift less than 2 min arc/hr. Simultaneous position and velocity recordings were made for each eye both during viewing and when under cover. The patients were seated in a chair with a head brace and chin rest at the center of a 5 ft radius arc containing red light-emitting diodes. Recordings were carried out in subdued light and calibration was accomplished monocularly for each eye (the other eye being behind cover). In the early 1970’s our studies of foveation in congenital forms of nystagmus, using laser-target retinal cinematography, revealed that, for both CN and LN, the slow phases brought the fovea off target and the fast phases returned it to the target (or, for some CN waveforms, towards the target). This relation of LN waveform to foveation was discussed in Daroff et al. (1978), shown in Fig. 10 of Dell’Osso et al. (1979) and has been observed by Lang (1978). With this knowledge we were able to align exactly the tracings for each fixating eye and, when the cover was removed, could detect and had a record of even the smallest tropias during binocular viewing. The term, ‘binocular viewing’ is used to describe the condition where both eyes are open and does not imply ‘binocular fixation’. After calibration, the patients were asked to view each light-emitting diode as it came on and a record was made of the ongoing nystagmus at all gaze angles of interest. Then, a near target was used to induce convergence and the eye movements were recorded both during the act of convergence and the return to distance viewing.

SUBJECTS

Of the three subjects of this report, Case 1 was sent to us with the clinical diagnosis of strabismus and LN, Case 2 with ‘acquired nystagmus’ since she appeared to have nystagmus only in lateral gaze, and Case 3 with strabismus and ‘nystagmus — presumably congenital’. Detailed clinical histories were unavailable to us.

RESULTS

The oculographic records of Case 1 shown in Figs. 1 and 2 document the absence of nystagmus during binocular viewing and its presence upon covering of either eye. In Fig. 1, the nystagmus commenced with cover of the left eye (at first arrow) and continued until the left eye was uncovered (at second arrow). It is evident from the right eye tracings that this eye continued to fixate the target under both conditions since the fast phases of the nystagmus brought the eye to the same position it held prior to covering the left eye (i.e., on target). However, it is
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Fig. 1. Position (pos) and velocity (vel) tracings of the right (OD) and left (OS) eyes during covering (left arrows) and uncovering (right arrows) of the left eye while viewing a target in primary position (Case I). Primary position is indicated by the horizontal line labeled 0°. Eye movement directions — right (R) and left (L) are indicated as are blinks (b). The symbols used in this figure are also used in Figs. 2 and 3.

equally evident from the left eye tracings that, once covered, the eye moved to a 15 deg eso position which it maintained during the ongoing latent nystagmus. The left eye did not return to the target until the cover was removed at the second arrow. Thus, when both eyes were uncovered, no strabismus was present and upon covering the left eye it began its esophoric movement simultaneously with onset of the LN. Fig. 2 shows the analogous results obtained by covering the right eye. Again, with binocular viewing there was no nystagmus; covering of the right eye (at first arrow) produced both a 15 deg esophoria of the right eye and jerk left LN. Uncovering of the right eye resulted in a cessation of the jerk left LN and resumption of binocular, aligned viewing. The square wave jerks seen during this latter period of binocular viewing and in Fig. 3 are not abnormal. Note that the left eye remained fixated on the target throughout the time period shown in Fig. 2. The observations documented in Figs. 1 and 2 are representative of all three patients. That is, all patients were recorded during period of binocular alignment with no nystagmus and during cover-induced phoria with nystagmus (LN). Thus, they all exhibited the ocular motor signs of pure LN and they all developed strabismus (i.e., a phoria) when either eye was covered.

Fig. 3 provides the critical link between MLN and LN. Initially, with both eyes open and viewing a target in primary position, there was a jerk right MLN, the right eye
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Fig. 2. Eye position and velocity of the right and left eyes during covering (left arrows) and uncovering (right arrows) of the right eye while viewing a target in primary position (Case 1).

was fixating the target, and the left eye was in an esotropic position (approximately 15 deg). As the left eye moved spontaneously from its esotropic position to the straight position, the jerk right MLN ceased. As long as the eyes remained aligned, there was no nystagmus. This sequence of events occurred repeatedly throughout the recording of this patient as well as those of the other two patients. They all exhibited periods of MLN accompanied by manifest strabismus.

Since no nystagmus was present when both eyes were open and aligned (the usual condition for these particular patients) and convergence on a near target did not elicit nystagmus, there was no effect of convergence on nystagmus.

Fig. 3. Position and velocity tracings of the right and left eye while binocularly viewing a target in primary position. Initially, the right eye was fixating the target and the left eye was esotropic. As the left eye tropia spontaneously was reduced to zero, resulting in both eyes being aligned on the target, the nystagmus ceased (Case 1).
DISCUSSION

The first description of latent nystagmus (LN) was by Faucon in 1872. Although several similar cases were described by others in the ensuing years, it was not until 1912 that C. & H. Fromaget described a case under the name 'latent nystagmus'. This term resulted from an analogy with latent squint. The Fromagets eventually reported ten cases of LN which included cases where:

- no nystagmus was present when both eyes were open (even in the extreme positions);
- nystagmus was present with both eyes open if the eyes were either converging or in extreme positions; and
- cases where nystagmus was present normally with both eyes open (i.e., manifest latent nystagmus — MLN).

Despite this wide variability of the signs of LN, the one feature that all cases had in common was that the occlusion of an eye either converted a latent nystagmus (i.e., no nystagmus) into one which was manifest or made the manifest nystagmus more marked. An excellent review of the early work on latent nystagmus was written by Sorsby in 1931.

It is noteworthy that the original case reported by Faucon (1872) was one in which the nystagmus was usually, though not always, present with both eyes open (i.e., he actually described a patient with both LN and MLN). It is important to emphasize this in light of the common misconception that LN is only present upon occlusion of one eye. The use of the term 'occlusion nystagmus' is misleading and serves to perpetuate this misunderstanding. The term 'latent' is used here with respect to strabismus and not to occlusion; that is, as soon as a tropia or phoria begins to develop the nystagmus commences. In this regard, the clinical presentation of our Case 2 should have raised suspicions of LN and, had this diagnosis been confirmed by ocular motility tests sooner, the more invasive neuroradiological test to which she was subjected could have been avoided. It is our experience that latent nystagmus in which there is absolutely no nystagmus with both eyes open in all fields of gaze is extremely rare.

More common is the case where the nystagmus appears in lateral gaze with both eyes open (as was our Case 2). It is this type which mimics gaze-evoked nystagmus and can lead the clinician astray. Most common are the cases where the LN is actually MLN since there is nystagmus with both eyes open although only one eye is fixating, the other being cortically suppressed.

Before discussing the results of this study, it is important to review the methodology employed. As required for any good ocular motility study, the recording system had low drift (to determine accurately eye position), low-noise (to distinguish small eye movements), and high sensitivity (to measure small tropias and phorias). Given the proper recording equipment, it was imperative that calibration be made during monocular viewing (i.e., one eye had to be behind cover). It is only when one can be sure that a specific eye is fixating the primary-position target that the position tracing of that eye can be properly aligned to the zero line on the recording paper. This is true whether or not there is an ongoing nystagmus in either or both eyes and whether there is normal (NRC) or abnormal (ARC) retinal correspondence. When each eye is properly calibrated monocularly, cover can be removed and the presence or absence of small tropias easily determined and measured objectively. Similarly, when
either eye is placed under cover, small phorias can be measured since both eyes are being simultaneously recorded whether or not they can see the target. Accurate, quantitative oculography provides an objective method to measure both the magnitudes of tropias and phorias in patients with nystagmus and to record their variation with time; the latter is impossible with subjective clinical office procedures which are difficult and often yield ambiguous results. We have employed this method of calibration for many years whenever recording patients with congenital varieties of nystagmus (since a strong possibility of strabismus exists). We also routinely calibrate in this way whenever we are interested in exact eye position of both eyes simultaneously; this applies to studies of normals as well as patients with ocular motor signs. This calibration procedure was pivotal in obtaining the insights into mechanism and relationship to strabismus of LN, in this study, and MLN, in our previous study (Dell’Osso et al., 1979).

Figs. 1 and 2 document, in a patient with true LN, the relationship between the two eyes (i.e., their position vis-à-vis the target) during the conditions when the eyes are motionless (binocular viewing) and when there is nystagmus (one eye occluded). Also shown for each eye are the transitions between binocular viewing and cover, and vice-versa. Clearly shown are the simultaneous beginning of a phoria in the covered eye and latent nystagmus in both eyes; also shown is the simultaneous return to alignment and disappearance of nystagmus when cover is removed. These findings are identical to those found for patients with MLN when the viewing eye was fixating the target and the eye behind cover was either eso or exo (Dell’Osso et al., 1979). Fig. 3 shows the presence of an esotropia during a brief period when this patient exhibited MLN and the subsequent simultaneous cessation of nystagmus and return to binocular alignment. Thus, for both MLN and LN, the presence of nystagmus was always accompanied by a mis-alignment (strabismus) of the nonviewing eye, be it behind cover (LN and MLN) or cortically suppressed (MLN).

Our hypothesized relationship between strabismus and latent or manifest latent nystagmus is summarized in Table 1. Since the patients referred to our laboratory for ocular motility studies had not been tested for retinal correspondence, the statements made below which address this point are based on the reported observation of others and our attempt to incorporate their findings into one consistent hypothesis. LN and MLN result in nystagmus of both eyes when one eye is covered; simultaneous with the nystagmus is the development of a phoria. When both eyes are open and aligned, no nystagmus exists if the patient has true LN and NRC. If MLN exists with both eyes open and NRC, a tropia will accompany it; with ARC, no nystagmus may result (i.e., the patient may exhibit pure LN). If the patient has both true LN and, at times, MLN (as did the three patients we studied), under the condition where both eyes are open, when there is no nystagmus, the eyes will be aligned and when there is MLN, either eye will assume a tropic position. In all cases of LN or MLN uncontaminated by the presence of CN, the nystagmus will have a jerk waveform whose fast phases are in the direction of the uncovered or fixating eye and whose slow phases are of decreasing velocity. We have
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### TABLE 1

<table>
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<tr>
<th>DX</th>
<th>Strabismus</th>
<th>One eye covered</th>
<th>Nyst.*</th>
<th>Both eyes open</th>
<th>Nyst.*</th>
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</thead>
<tbody>
<tr>
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<td>Yes</td>
<td>Phoria</td>
<td>Yes</td>
<td>Orthotropic (NRC)</td>
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<td></td>
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<td>Yes</td>
<td>Tropia (NRC)</td>
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<tr>
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<td>Yes</td>
<td>Orthotropic (NRC)</td>
<td>No</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Tropia (NRC)</td>
<td>Yes</td>
</tr>
</tbody>
</table>

* In all cases the nystagmus is a jerk nystagmus with fast phases in the direction of the uncovered or viewing eye and decreasing-velocity slow phases.

DX — Diagnosis; Nyst. — Nystagmus; NRC — Normal retinal correspondence; ARC — Abnormal retinal correspondence.

recorded patients with both LN/MLN and CN who have exhibited complex combinations of the waveforms of both as well as each type individually; discussion of such patients is beyond the scope of this report.

Based on our observations in these three LN cases and those made in our previous study of 31 MLN cases, the following relationship to strabismus emerges: Strabismus, as we use the term, is a necessary condition for both LN and MLN. That is, the presence of LN or MLN implies the existence of a phoria behind cover and further, MLN implies the existence of a tropia during binocular viewing. Also, we feel that it is possible for a patient with a tropia to have ARC and true LN during binocular viewing (i.e., no manifest nystagmus). Since there are patients with strabismus who have neither LN nor MLN, it is obvious that, although strabismus is a necessary condition for both, it is not a sufficient condition for either. Consistent with our hypothesis that strabismus is necessary for LN and MLN is the observation first made by Healy (1952) and repeated a decade later (Healy, 1962) that, when an amblyoscope was used to move the two images onto the foveas of a patient with a tropia, NRC and nystagmus (MLN), the nystagmus stopped when fusion was achieved. This is analogous to the absence of nystagmus in our patients only when the eyes were aligned; since proper alignment with no LN was the common condition in our patients and our recordings showed their tropias to be intermittent, small, and variable, it is highly probable that they had NRC.

Two recent papers claim that LN can exist (albeit rarely) in patients who do not have strabismus. Lang (1978) found strabismus in 99% (196 patients out of 198) of patients he diagnosed as having LN. Thus, two patients apparently had no strabismus. However, both the LN and the strabismus were diagnosed clinically without accurate ocular motility recordings. Lang admits that these ‘LN’ patients could also show ‘spontaneous’ nystagmus and that it is a ‘great curiosity’ to find LN without strabismus. Given these facts, we contend that his findings actually support our hypothesis that all patients with proven LN (i.e., proven by accurate ocular motility recordings) have strabismus; it is simply impossible to
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clinically differentiate some cases of CN with a superimposed latent component from MLN and it is quite possible that such patients were included in Lang's sample. Alternatively, in the presence of nystagmus a small tropia could easily have gone undetected (see below). A second study by Ishikawa (1979) does contain some eye movement recordings. His claim that five patients with 'orthophoria' had LN is, in our opinion, explicitly contradicted by his Fig. 2. Upon covering of the right eye in an 'orthophoric' patient, the left eye had to move 1 deg in an eso direction to take up fixation; jerk-left LN is shown during this interval, confirming fixation with the left eye. When cover was removed the left eye returned to its original position. From this eye movement recording we can only conclude that this patient actually had a 1 deg exotropia; the tracings are inconsistent with our understanding of the term 'orthophoria'. It is probable that such small tropias may be impossible to detect clinically in the presence of LN; once shown by objective recordings however, they cannot be ignored in favor of the subjective clinical impression of orthophoria. Thus, we also cite Ishikawa's data as supporting our contention that strabismus is a necessary accompaniment to LN.

The mechanism for latent and manifest latent nystagmus, which we briefly presented in 1978 (Daroff et al., 1978) and more fully discussed in two later publications (Dell'Osso et al., 1979; Dell'Osso & Daroff, 1981), remains the only one which we find tenable and consistent with both our observations and the data of others. That is, the direction of the conjugate slow phase exponential drift in LN and MLN is always nasal to the viewing eye and is independent of gaze angle, strabismus angle or tropia type. Our hypothesis was based on the fact that perceived egocentric direction is calculated differently under binocular and monocular viewing conditions. We postulated that patients with LN and/or MLN are unable to correctly switch between these two mechanisms and that their inability to do so results in a conjugate nasal exponential drift towards the improperly perceived version angle. With this new evidence on the necessity of strabismus, we now suggest that the combination of strabismus and the inability to shift between the perceived egocentric directions associated with both binocular and monocular vision is necessary and sufficient for the development of latent and/or manifest latent nystagmus.

Given out observation that the onsets of any tropias or phorias were simultaneous with the onset of nystagmus, one might not expect to see recordings of an LN/MLN patient showing the absence of nystagmus during the presence of a tropia. Although this expectation does not follow logically from our assertion that strabismus is necessary for LN/MLN, it is fulfilled for most cases we have studied. None of the three cases presented here nor 30 of the 31 MLN cases we previously studied showed a tropia in the absence of MLN. However, one of our MLN patients with unidirectional MLN (left beating) did have a slight left esotropia when fixating with the right eye; no MLN was present under these conditions (Fig. 6 in Dell'Osso et al., 1979). Also, Ciuffreda (1977) described a patient in his dissertation who was said to have pure LN and a constant small-angle right esotropia of 6 prism diopters. If her binocular viewing records showed no nystagmus (as
was stated in the dissertation) and the development of LN only upon covering of either eye, then it would at first seem that both she and the patient with unidirectional MLN we studied represent a contradiction of our hypothesis. In fact they do not; strabismus is necessary but not sufficient for LN/MLN and even in patients with LN or MLN there can exist conditions when the strabismus is manifest but the nystagmus is not. A possible explanation for this occurrence is that these patients, unlike the three patients of this report, had non-variable, small-angle esotropias; such patients very frequently have ARC (Burian & Von Noorden, 1974). In cases like these, the existence of ARC may be analogous to binocular alignment vis-à-vis the development of MLN when both eyes are open with the tropic eye in its normal, slightly esotropic position. Until more data is accumulated from this class of patients, the relationship of ARC to LN remains speculative. For those patients with intermittent, variable esotropias, normal retinal correspondence and LN or MLN, we have found the onset of a phoria or tropia to be simultaneous with the onset of the nystagmus.

Although the subject described as Case 1 in Dr. Ciuffreda’s dissertation had pure LN at the time she was studied (age 18), her history revealed that she had both esotropia and ‘manifest nystagmus’ which were noted shortly after birth. After strabismus surgery performed by Dr. William Hoyt when the patient was one year old, the tropia was corrected and the ‘manifest nystagmus’ reduced. When recorded by Dr. Ciuffreda, she had no nystagmus under binocular viewing conditions and only developed LN upon covering of either eye. Based upon our experience in recording more than 350 patients with congenital types of nystagmus, we believe that the ‘manifest nystagmus’ noted shortly after birth was actually MLN since there is no evidence that strabismus surgery could convert CN to LN; their mechanisms are entirely different. The effect of Healy’s moving the image to one eye and overcoming a fixed tropia (Healy, 1952, 1962) was similar to that which we observed for variable tropia when the eye itself became aligned with the fixating eye; in both instances MLN ceases. Thus, it is quite possible that young children born with fixed tropias and MLN can benefit greatly by early strabismus surgery. If their eye can be aligned and their MLN eliminated, such patients may be left with only pure LN which, under normal binocular viewing conditions, would not affect their visual acuities. To our knowledge this is the first report of the conversion of MLN (inferred but not documented) to LN by strabismus surgery where the LN (i.e., the absence of nystagmus under binocular viewing conditions) had been documented by accurate eye movement recording (Ciuffreda, 1977). The clinical implications of this speculation are obvious and warrant further study to determine whether it is in the best interest of such patients’ future acuities to perform strabismus surgery as early as possible when manifest latent nystagmus co-exists with a fixed tropia.

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