18

Congenital and Other Types of Infantile Nystagmus: Recording, Diagnosis, and Treatment

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There are several types of benign nystagmus that may appear at birth or in early infancy. All these specific types belong in the general category of *nystagmus of infancy (infantile nystagmus)* and are caused by different abnormalities in the ocular motor system. They exhibit different waveforms that reflect these specific abnormalities and also have individual names to reflect the differences (waveform, mechanism, and clinical) among them. In addition, infants may exhibit other, more malignant types of nystagmus, which suggest neurologic disorders and require immediate attention.

Congenital nystagmus (CN) is a specific type of infantile nystagmus that usually appears in early infancy. There are other types of nystagmus of infancy that are not CN and should not be grouped together with CN under the term *congenital nystagmus*. The word "congenital" means "with birth," and although CN may be present at birth, it usually becomes manifest in early infancy, and it may not become manifest until later in infancy, childhood, or even adulthood (1). Therefore CN should be thought of as a description of a congenital predisposition toward a specific ocular motor instability that may be manifest at birth, in early infancy, or, occasionally, later in life. Attempts to change the name of this specific motor sign to one that merely shifts the time of onset (such as "infantile" nystagmus) are pointless, since the time of onset is a variable with a wide range in individuals. If the name is to be changed at all, and I am not advocating that it should be, the best choice should reflect the waveform(s) that are intimately related to the underlying mechanism(s) instead of some arbitrary time of onset. The latter only serves to exclude those with the same disorder but different timing. Marshall M. Parks (discussing "congenital" vs "infantile" esotropia) said, "To adopt another term in place of the widely used congenital esotropia, just because the esotropia cannot be proven to be present at birth may not be in the best interest of strabismology" (2). The same applies to CN and its study, since the most important factor in the treatment of CN is not its name but its accurate diagnosis and differentiation from other benign types of nystagmus of infancy and from malignant acquired types.

RECORDING SYSTEM CRITERIA

Accurate diagnosis of CN and the other types of nystagmus of infancy is dependent on waveform analysis of the oscillations of

both eyes. Therefore, it is important that the recording system chosen for the task meets certain minimal criteria regarding the recording of both eyes simultaneously, sensitivity, freedom from noise and drift, electronic signal conditioning, and freedom from distortion of the nystagmus waveforms. These criteria must be met by whatever method is used if the diagnostic or therapeutic conclusions drawn from eye movement data are to be an improvement on clinical impressions. Confusion between CN and other types of infantile nystagmus can result if only clinical impressions or distorted renditions of the nystagmus that do not show the movements of both eyes accurately and simultaneously are used.

There are many articles and review chapters in the literature covering each method currently in use and listing the underlying principles of operation, advantages, and limitations. In this chapter, I concentrate on the criteria that must be satisfied to produce data that can be used for diagnosis and treatment of nystagmus. There are conditions when each of the available methods will suffice. Determination of those conditions requires an understanding of the measurement criteria needed for a particular study. We can divide the eye movement recording criteria that apply to the study of nystagmus into two sets: research and clinical.

Research Criteria

The criteria for research into the mechanisms of nystagmus or other eye movement abnormalities are more stringent than those needed in the clinic for diagnosis and treatment. They are summarized as follows.

1. The simultaneous recording of *both* eyes is required. Conjugacy should not be assumed (indeed, in a patient population, conjugacy is not the rule), and comparison of the oscillations of the two eyes is important for differentiation between some types of nystagmus.

2. The system must be DC-coupled; that is, accurate position information must be provided. AC-coupling results in lost position information and distortion of slow phase waveforms. Both can prevent accurate identification of the type of nystagmus being recorded.
3. The system bandwidth should be about 100 Hz to allow accurate recording of saccades and higher-frequency components of some nystagmus waveforms.
4. The system sensitivity should be high enough and noise low enough to detect small movements (i.e., several minarc).
5. There should be little or no drift over the time frame of all recording sessions.
6. The system should be as noninvasive as possible to allow for longer studies and a larger population of subjects and to prevent the system from introducing unnatural eye movement behavior.
7. The system should be able to record eye movements with enough range in all three planes (horizontal, vertical, and torsional) if possible. This allows the maximum flexibility in studying many types of eye movements.
8. The system should be insensitive to translational head movements, or head stabilization must be provided.
9. Primary position data must be differentiated (analog or digital differentiation) to obtain eye velocities. The bandwidth of the differentiator should be about 100 Hz for saccades and 20 Hz for slow phases and smooth pursuit.

These criteria rule out electrooculography (EOG) as an effective research tool except under narrowly defined circumstances. Its continued use reflects its ease of application, but EOG usually provides data that are unsatisfactory (i.e., too much noise, too little sensitivity and accuracy) for use in research. The use of EOG is further hampered by the common use of equipment and methods designed for electronystagmography instead of ocular motility studies. Electronystagmography com-

monly uses bitemporal electrode placement (yielding no true data about either eye) and AC-coupling (eliminating position information and distorting slow phases). Both of these practices are unacceptable for analysis of all types of infantile nystagmus. Head-mounted infrared reflection (IR) and other reflection methods satisfy many of the criteria, are most useful in the horizontal plane (±20–30 degrees), are of limited use in the vertical plane (±10 degrees), and cannot detect torsional eye movements. The magnetic scleral search coil system is the most accurate and can be used in all planes but is invasive, precluding its use in some patients.

Clinical Criteria

In the clinic, some of the listed criteria may be relaxed slightly. The following are guidelines for recording systems used in the clinical evaluation of nystagmus.

1. and 2. Same as for research criteria.
3. The system bandwidth should be about 50 Hz unless accurate recording of saccades and higher-frequency components of some nystagmus waveforms are required.
4. The system sensitivity should be high enough and noise low enough to detect movements of about 0.5 to 1 degrees.
5. There should be little or no drift over the time frame of all recording sessions, or frequent recalibration will be necessary.
6. Same as for research criteria.
7. The system should be able to record eye movements with enough range in two of the three planes (horizontal and vertical) if possible. That allows for flexibility in studying the common types of eye movement disorders.
8. The system should be insensitive to translational head movements, or some head stabilization must be provided. For infants, the head can be adequately stabilized by an assistant placing his or her hands on either side of the subject's head. This will allow waveforms to be recorded and diagnoses made.
9. Primary position data should be differentiated (analog or digital differentiation) to obtain eye velocities. The bandwidth of the differentiator should be about 20 Hz unless saccadic velocities are being studied.

These criteria allow for the use of EOG only if the question under consideration can be answered within the limits of the criteria. It is possible to record the waveforms of nystagmus in each eye if the amplitude is great enough. The large horizontal range of EOG allows recordings at lateral gaze angles up to 30 degrees. Bitemporal electrode placement and AC-coupling are unacceptable for clinical diagnosis or treatment. At best, bitemporal EOG provides an approximation of nystagmus waveforms of a "cyclopean eye," reflecting the vector sum of the movements of the two eyes. It can be a grossly misleading rendition of the movements of either eye. IR is most useful in studies of horizontal and vertical nystagmus, the latter near primary position. Mounting the sensors on eyeglass frames reduces the sensitivity to head movements. IR and EOG recording methods are easy to use (after minimal training) and noninvasive. The magnetic scleral search coil system can be used in a clinical setting but is invasive (and expensive).

Calibration Techniques

Adults and Children

Proper calibration of each eye individually is necessary to accurately measure a null angle or tropia. The subject should be seated with the head restrained in the primary position. Zero should be set by adjusting the foveation periods of the CN subject's waveform to the zero position while the subject is fixating a target at 0 degrees. This must be done for each eye while the

other is occluded. Once done, both phorias and tropias of the nonfixing eye will be measurable from the records if either are present. Sometimes, the foveation periods are not easily determined by their long duration. They occur at the end of the fast phases of jerk CN and during the more flattened peaks of pendular CN. For subjects with latent/manifest latent nystagmus (LMLN), the zero position is at the end of the fast phases of the nystagmus or, if dynamic overshoots are present on the fast phases, at their termination. After zero is set for an eye, the gain should be adjusted to place the foveation periods at the proper displacement from primary depending on the calibration targets used (e.g., ± 15 or 20 degrees). In CN, the direction of the nystagmus may reverse as gaze is directed from one target to the other. In LMLN, the direction cannot reverse if one eye is occluded (Fig. 1). Once calibrated, the nystagmus amplitudes and positions of each

eye can be recorded for both monocular and binocular (if possible for the subject being tested) fixation.

Infants

Unfortunately, due to the limitations of current eye movement measurement devices, is not possible to accurately calibrate infants. However, accurate calibration is not needed for diagnostic purposes. The nystagmus waveform as well as the phase relationship between the nystagmus of the two eyes can be obtained without calibration. When using an IR device, the structure holding the emitters and phototransistors is removed from the eyeglass frames, and two hands are placed against the infant's temples to steady the structure in front of the infant. With the correct interpupillary distance of the recording sensors preset, accurate recordings from both eyes
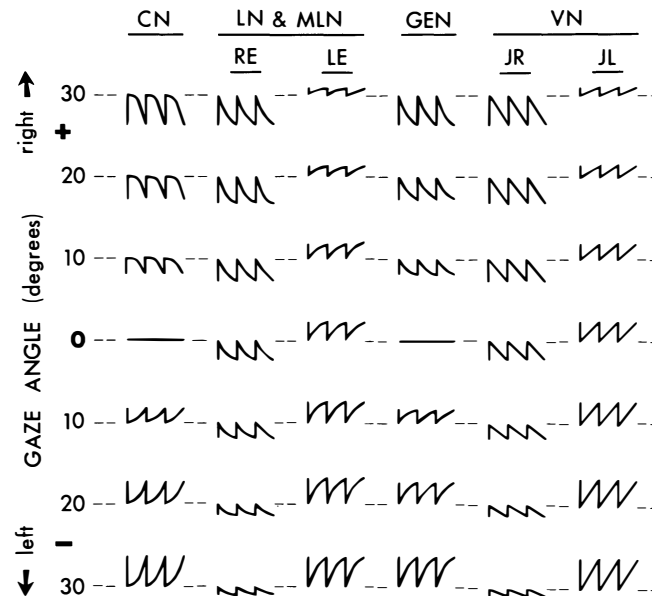

FIG. 1. Illustration of eye movement recordings of congenital (CN), latent (LN), manifest latent (MLN), gaze-evoked (GEN), and vestibular (VN) nystagmus. Three beats of each type of nystagmus at several gaze angles are shown. RE and LE indicate the viewing eye, and JR (jerk right) and JL (jerk left) indicate the nystagmus direction. Note the differences in the slow phases, variation over gaze angle, and the fact that the fast phases are foveating. (From ref. 36.)

with minimal head movement artifact can be obtained. The head is easily held still by the experimenter who is holding the sensors in position. Approximate calibration is possible by getting the infant's attention to targets at ±15 degrees. This technique has been applied successfully in patients ranging in age from premature infancy (while still in an incubator) to 2 years.

RELATION TO STRABISMUS

Since there is a high incidence of strabismus in individuals with different types of infantile nystagmus, it is important to understand the relation of strabismus to nystagmus, that is, to CN or LMLN. Of all strabismus patients, about 50% will not have any type of nystagmus. The other 50% will have nystagmus, and the question is, which type? Of that 50% of the total, 26.5% have CN, 17% have LMLN, and 6% have both CN and LMLN. The presence of LMLN means there is an obligate strabismus. Of 100 consecutively seen patients with both nystagmus and strabismus, CN is more probable than LMLN because there are many more CN patients. This is true despite the fact that all patients with LMLN have strabismus. For 100 consecutively seen patients with CN, LMLN, or both, we found that 43% had strabismus (3). Therefore, a patient with nystagmus will more often not have strabismus. If one considers only CN patients (including those with some LMLN), almost 75% will not have strabismus. Thus, most CN patients do not have strabismus. For those with pure CN, the percentage will be even higher. All 12 CN waveforms have been recorded in patients with binocular alignment, indicating that it is not necessary to have strabismus for any particular CN waveform. Of the 43% of patients with strabismus in the first 100 patients mentioned, more than half had CN. This finding is of interest when one considers that most patients with CN do not have strabismus. Summarizing, for patients with strabismus and nystagmus, 53% will have CN, 35% will have LMLN, and 12% will have both CN and LMLN.

It is not known if infants with spasmus nutans have strabismus. However, since their eyes oscillate out of phase with each other, these patients probably experience oscillating diplopia (oscillopsia and variable diplopia).

CONGENITAL NYSTAGMUS

Differential Diagnosis

As stated previously, in addition to CN, other types of nystagmus (and saccadic oscillations) may occur at or shortly after birth and should not be confused with CN. The other types of infantile nystagmus are different from CN in waveform (mechanism) and clinical characteristics. The other benign types of infantile nystagmus are LMLN, spasmus nutans, and the nystagmus blockage syndrome (NBS). The physician need not be concerned with whether the nystagmus appeared at birth or in the first few weeks after birth. More important is (a) determining whether this is a benign nystagmus or one that suggests disease and (b) if benign, determining whether it is CN, LMLN, spasmus nutans, or NBS. The types of nystagmus that are malignant are downbeat nystagmus (owing to structural brainstem abnormalities), epileptic nystagmus, uniocular nystagmus (owing to optic nerve glioma), vestibular nystagmus (owing to any of several vestibular problems), and "nystagmus" of the blind; the latter wandering eye movements are not truly nystagmus. Table 1 lists both benign and malignant types of infantile nystagmus.

It is important to distinguish between a benign and a malignant nystagmus when an adult is first seen after trauma or for some unrelated condition and a correct history is unavailable. Under these conditions, it may be difficult to determine if the nystagmus is a critical indication of the trauma or other

TABLE 1. *Types of infantile nystagmus*

Benign	Malignant
Congenital (CN)	Downbeat (structural
Latent/manifest latent	brainstem abnormalities)
(LMLN)	Epileptic
Spasmus nutans	Uniocular (optic nerve
Nystagmus blockage	glioma)
syndrome (NBS)	Vestibular
	"Nystagmus" of the blind[a]

[a]Wandering eye movements, *not* true nystagmus.

condition. Ocular motility analysis can identify CN and LMLN, preventing otherwise necessary invasive neurologic testing (e.g., angiography, lumbar puncture) that exposes the patient to morbidity and mortality risks and pain. When motility examination indicates an acquired nystagmus, recording the specific type of nystagmus is essential for neurologic diagnosis and treatment.

The *motor eye sign,* CN, is defined as follows: either a pendular or jerk nystagmus resulting from a slow eye movement instability, producing periodic motion of the eyes away from an intended gaze angle or target and back to the target (not across the target) (4). The pendular CN waveforms look sinusoidal but usually are distorted by both flattening and the presence of small foveating saccades on the peaks corresponding to where target foveation occurs. The jerk waveforms of CN are caused by an instability that leads to an acceleration of the eyes away from the intended gaze angle or target and requires a saccade (braking saccade) in the opposite direction to stop the runaway. This saccade might return the eyes back to the target (foveating saccade) or begin a slow eye movement back to the target for refoveation. The direction of CN is defined by the direction of the saccade, although it is the slow eye movement that causes the runaway. This is consistent with the convention used to define the direction of all types of nystagmus. CN is predominantly horizontal, with occasional vertical components in some patients. Torsional

components are common, and predominantly vertical CN is rare.

Sensory Defects

The following sensory defects are commonly associated with CN of all waveforms.

Achromatopsia
Albinism
Aniridia
Congenital stationary night blindness
Disorders of the media (congenital cateracts)
Optic atrophy (developmental/hereditary)
Optic nerve disease (aplasia/dysplasia/hypoplasia/atrophy/colobomas)

It is incorrect to equate pendular CN with a sensory defect or jerk CN with a motor defect. All CN waveforms are caused by a primary motor defect. Systematic analysis of eye movement recordings has shown that there are no specific waveforms associated with sensory defects. The etiology and the mechanisms underlying CN waveforms are independent of accompanying sensory defects (5). Therefore, although there is an association between many sensory defects and the presence of CN (6), it is not a primary, causal relationship. The sensory defect is not the direct cause of the nystagmus, nor is the nystagmus the direct cause of the defect. Sensory defects appear to provoke the manifestation of CN in individuals in whom there is a predisposition for CN. Thus, although sensory defects are not the primary cause of CN, they appear to be the precipitating factors in the manifestation of CN in some patients. Despite the eye movement data that proves conclusively that there is only one CN (i.e., a motor sign indicating ocular motor instability), the early, erroneous idea that there existed two types of CN, "sensory" and "motor," persists to this day (6). The term "motor CN" is redundant, and "sensory CN" is incorrect for the reasons discussed. Neither

term should be used. Some investigators have claimed that eye movement data "cannot differentiate" between these two putative types of CN (7). That is a naive interpretation of the available evidence. Since sensory defects do exist in patients without CN and CN exists in many patients who do not have any sensory defects, simple logic dictates that sensory defects cannot be either the necessary or sufficient condition for the development of CN.

Since any of several different sensory abnormalities (each affecting the primary visual signal in unrelated ways) can result in the developing motor system becoming unstable, the following possibilities are suggested: (i) a small percentage of individuals are born with a motor system that is precariously close to oscillation, or (ii) the ocular motor system has evolved in such a way that the horizontal system is close to instability in many individuals. The logic for the latter hypothesis is based on the adaptive necessity for rapid horizontal eye motion to survive. Whatever the underlying reason, the motor system requires early visual input to assure its stability, and any sensory defect that compromises acquisition of that early visual input might also interfere with the development of motor stability in some. In those individuals with CN and no sensory defects, the motor system may have developed with an instability (owing to hereditary or spontaneous mutation) that persists in the presence of adequate visual input. The individuals in whom CN is truly congenital include both those with idiopathic CN and those with associated sensory defects, not just the former.

In individuals with a sensory defect, it, instead of the nystagmus, may be the limiting factor preventing good visual acuity, and in some of them, halting the eye motion may not result in an acuity improvement. In others (and in those without sensory defects), visual acuity can be improved by damping the CN. One must perform the necessary diagnostic tests (e.g., the electroretinogram, ERG) to correctly assess the functional integrity within the visual system and diagnose any sensory abnormalities present. However, neither this analysis nor a sensory diagnosis is sufficient to describe the motor defect causing the nystagmus. The latter requires motility recordings for diagnosis, since patients with sensory defects might have one of the other types of infantile nystagmus and not CN. Also, recordings are needed to identify the existence of gaze angle and convergence nulls in the treatment of CN. If a child with nystagmus has a best-corrected visual acuity of 20/60 (or an adult has 20/50), one should look carefully for afferent defects. Since young children with CN are more often examined than adults, sensory defects may seem to be more prominent in pediatric patient populations than in adult populations (6). At present, there is insufficient evidence to link CN and sensory defects to one gene or to different genes with variable probabilities of interaction. Further speculation in this area is outside the scope of this chapter. The diagnosis and treatment of the various sensory abnormalities that may accompany CN are not covered in this chapter, which concentrates on the proper management of CN and other forms of infantile nystagmus.

Diagnostic Criteria

The diagnostic characteristics for each of the benign types of nystagmus of infancy are unambiguous. Each type can be differentiated by simple eye movement recordings that simultaneously provide the data necessary for effective therapeutic intervention.

Waveform

Regardless of time of onset, the waveforms recorded from individuals with CN are identifiable. Since the waveforms are reliably identifiable over time, the motor instability in all instances is constant, indicat-

TABLE 2. *Waveforms of benign types of infantile nystagmus*

Congenital[a]	Latent	Spasmus nutans
Pendular		Pendular (variable conjugacy)
Pendular		
Asymmetric pendular		
Pendular with R/L[b] foveating saccades		
Unidirectional jerk[c]		
Jerk R/L	Jerk R/L	
Jerk R/L with extended foveation		
Pseudocycloid R/L		
Pseudojerk R/L		
Bidirectional jerk[c]		
Pseudopendular		
Pseudopendular with R/L foveating saccades		
Triangular		
Bidirectional jerk R/L		
Dual[c]		
Dual jerk R/L	Dual jerk R/L latent	

[a]The nystagmus blockage syndrome exhibits one or more of the congenital waveforms and may exhibit the latent waveform (see text).
[b]R, right; L, left.
[c]Accelerating slow phases in congenital waveforms and decelerating slow phases in the latent waveform.

ing that there is only one CN, independent of time of onset. With the aid of recorded eye movements, the diagnostic criteria for CN are definitive. Any of the 12 waveforms (Table 2) that have been identified in CN (8) are pathognomonic of CN except pure pendular, which could be an acquired nystagmus. However, the pendular nystagmus of CN is usually distorted, allowing for longer foveation periods, whereas acquired pendular nystagmus is not distorted. In acquired nystagmus, the slow phases can be linear, of increasing or decreasing velocity, as well as pendular. CN slow phase can be pendular, of increasing velocity, or some may look linear. In CN, we identified within the pendular or jerk major catagories, 3 different forms of pendular and 8 jerk (4 unidirectional and 4 bidirectional) waveforms. One waveform, dual jerk, is the superimposition of pendular on a jerk waveform with increasing velocity slow phases. Illustrations and examples of specific CN waveforms are available in the literature (8,9). Figure 1 illustrates a typical jerk CN waveform in comparison to LMLN and two types of acquired nystagmus. In a group of 100 patients with CN, 87% had various combinations of the 12 waveforms. The remaining 13% exhibited only 1 waveform. Distinguishing the subtle characteristics of CN waveforms requires DC-coupled, high-bandwidth recordings of both eyes simultaneously.

One specific group of patients who usually have CN are those with albinism. Just as eye movement recordings can diagnose CN, albinism can now be diagnosed definitively by a test based on the visual evoked potential (VEP). If performed properly using the paradigm developed by Apkarian and Spekeijse (10), the VEP is virtually 100% free of both false positives and false negatives. This paradigm uses full-field, monocular pattern (checkerboard) onset/offset stimuli instead of pattern reversal stimuli. The latter contain motion artifacts and should not be used in individuals with CN or any other nystagmus. For children below the age of about 3 years, reliable results are obtained with a luminance flash paradigm. The appropriate VEP paradigm demonstrates the unequivocal dissociation between the misrouted fibers of albinos and their CN, since no individuals with hereditary or idiopathic CN had misrouting.

Thus, the hypothesis that misrouting can cause CN is incorrect and lacks physiologic foundation (11).

Symptomology

In addition to the quantifiable and definitively diagnostic characteristics of CN available through eye movement recordings, there are significant clinical signs. In CN, one should look for a *null angle,* an indication of which is the presence of a head turn. A teenager or an adult may not show a head turn because of societal pressures. They have learned to keep their heads straight at the expense of good vision, since it is not appropriate to walk around with a turned head. However, children exhibit spontaneous head turns. A positive family history and negative neurologic examination also suggest CN.

If converging the eyes damps the CN, patients will hold reading material close. They may have a latent component, which is checked by a cover test. If the nystagmus direction reverses under cover, one still does not know whether the nystagmus is LMLN or CN with a latent component (they are different). Patients may exhibit head nodding. In CN, the head nodding is not compensatory.

Many patients with CN exhibit a *null angle of gaze* where the CN magnitude damps. The definition of a null (a true null) is that on either side of the damped position the nystagmus must increase in magnitude (Fig. 1). That is in contrast to LMLN, in which monotonic variation with gaze angle (Alexander's law) causes patients to keep their eyes deviated to one side where the nystagmus is low. LMLN does not have a true null, since there is no increase on both sides (Fig. 1). In CN, the position of the null is a function of the angle of gaze and also a function of the velocity of the eyes. (12,13). Thus, the null angle during fixation (static null) usually does not equal the null angle during pursuit, optokinetic nystag-

mus, or head movement, where the vestibulo-ocular reflex (VOR) is stimulated (dynamic null). Usually, the null is shifted in the direction opposite to the eye movement. During pursuit to the left, the dynamic null moves to the right of the static null, and during pursuit to the right, the dynamic null moves to the left. In a sample of 100 CN patients, 48% had both convergence and gaze angle nulls, 29% had only convergence nulls, 9% had only gaze angle nulls, and there were 14% with no nulls. Accurate measurement of the null position requires a well-calibrated, DC-coupled recording system. That measurement can then be used to prescribe prisms or determine the amount of surgery to be performed (see the section on Treatment).

Children with CN automatically adopt a head turn to see better. They do not have to wait to be told about the null angle. They know where it is because things appear clearer when they turn their head, placing their eyes at the static null angle. One may think of the null as a region of ocular motor equilibrium. The brain stem (left and right) generates forces pulling the eyes both ways, and there is a position of equilibrium of forces, not necessarily in primary position, where the nystagmus is minimal. When viewing targets at the null angle, acuity should increase. When there is a severe afferent defect, acuity will not increase measurably.

Sometimes a patient will tilt the head, sometimes turn it (vertically or horizontally), and simultaneously tilt it, suggesting that the oblique muscles may be involved. A few patients do have significant vertical, and many may have small torsional, components. Again, visual acuity increases with vertical or torsional head positions, especially if there are no afferent defects.

The mechanism causing a CN reversal when covering an eye is similar to when the person pursues. The null moves and can cause a direction reversal of the CN if gaze goes from one side of the null to the other (14). Although CN with a latent component looks like latent (LN) or manifest latent

(MLN) nystagmus, it is not, since the waveform remains CN. Only the CN direction changes because the null has moved with occlusion.

CN with a latent component can appear as though there were two null angles, since if the fixating eye changes with gaze angle, the induced null shift will appear to be a second null. In binocular individuals with CN, there is only one null, and in those with strabismus, there is only one null for fixation with either eye. A good recording system, properly calibrated for fixation with each eye, will detect shifts in the fixating eye (and the tropias of the nonfixating eye) and prevent misreading the records as showing two nulls. Also, CN and periodic (PAN) or aperiodic (APAN) alternating nystagmus will mimic two nulls because of the null shift accompanying the direction reversal. Steady fixation in primary position for several minutes will disclose PAN or APAN.

Many children with CN exhibit spontaneous head oscillations. Adults, when they are concentrating on a visual task (real or imaginary), also may exhibit head oscillations. These head oscillations use existing pathways in the neck muscles. EMG in the neck muscles show that when normals make saccades to the left, the innervation is seen in the left-turning neck muscles, and when they made them to the right, it is seen in the right-turning neck muscles. Normally, when we look left, we are going to turn our head left. The pathways exist, and if there is an instability causing the eyes to oscillate and one records from the neck muscles, the same waveform is seen. When the oscillation grows large enough, the head will start oscillating. This is not something willed by the patient to compensate for the CN (7). It is a manifestation of an existing oscillation on existing pathways to the neck. It has been thought that head oscillations were compensatory. The head was supposed to be moved equally and oppositely to the CN to stabilize the eye in space. If that were true, the VOR gain would have to be zero. A VOR is incompatible with a head movement that compensates for an eye movement. With no VOR, the head would have to move in complex ways opposite to the CN waveforms to achieve stability, which is clearly impossible. The head has too much mass to duplicate the waveforms of CN. The compensatory hypothesis is not valid when you understand the VOR. Realistic compensation could theoretically be accomplished if one could suppress a normal VOR to near zero and only move the head equally and oppositely to any movements of the eye during the CN foveation periods. This would achieve gaze stability during that part of the waveform and is a theoretically possible form of compensation useful only in those cases where the foveation periods of an individual were not stable with the head still.

In individuals with CN, the VOR is normal. It is not affected by CN (13,15). Therefore, the head oscillations of most people with CN are an extension of the CN, and during the foveation periods, the eyes do not move and acuity is unaffected by head movements (16–18). Basically, the head oscillations of an individual with CN and a normal VOR are equivalent to those of a normal person moving the head, and acuity does not change. For most CN patients (who have good foveation periods), there is no advantage to head nodding. If the foveation periods are flat, head motion cannot help the patient, and head nodding is not an adaptation designed for increasing acuity (7).

Treatment

In CN, a strong fixation reflex maintains foveation despite slow acceleration off the target. This is juxtaposed to the mistaken idea that CN is caused by poor fixation reflexes. We have found that fixation in CN is accurate (<13 min of arc) (19). In LMLN, the fixation reflexes appear to be ineffective, since as soon as a fast phase is

completed, the slow phase immediately takes the eyes away from the target. These patients seem to be unable to use their fixation system to keep the eye on the target. One can increase the visual acuity of CN patients (if they have no foveal defects) when there is a null angle or a convergence null and when there are good waveforms with long foveation periods. Then, prisms or surgery may be effective. For patients without these characteristics, these methods are not likely to be successful. One could still straighten the head, but an increase in acuity is unlikely. If one can minimize the attempt to fixate, acuity will be maximized, since in CN, the greater the effort to see (the fixation attempt), the higher the level of nystagmus. When a CN patient turns the head to use the null, the level of nystagmus at that null angle is higher than (a) if, via surgery, the null is moved to primary position, or (b) if, via prisms, the target is shifted to the null angle. These conditions are better, since there is no effort involved in seeing the target at the null angle.

Visual acuity is proportional to how much time in each cycle the target is foveated (20–23). The longer the eyes remain on the target, the more visual detail is obtainable. The retinal slip during this time should be less than 4 degrees/sec. Data from normal individuals show that when retinal slip stays below 4 degrees/sec, acuity is the greatest. Also important is the stability of the foveation periods. The eyes must be in exactly the same position from cycle to cycle; the extent to which that is not so is called *jitter*. Jitter must be minimized, otherwise even with long foveation periods, one cycle might fall to the left of the target and the next to the right. All these characteristics must be present for good acuity, that is, long foveation periods with low retinal slip velocities that are repeatedly positioned accurately on target. The nystagmus foveation function (NFF-duration of foveation period/product of the standard deviations of the mean position

and velocity of the foveation periods) is the best method for determining the conditions under which acuity will be maximum (19). It can be used to plan surgery, to prescribe optical correction, or for other therapy.

Optical Treatment

Prisms can be used to shift gaze and take advantage of nulls within 5 degrees of primary position. If there is a null to the left, base-right prisms will move the eyes leftward into the null. Accurate measurements of the gaze angle and convergence nulls can be translated directly to prisms. To converge the eyes, use base-out prisms with added minus to compensate for the induced accommodation. To move the eyes laterally and converge them, use compound prisms. Prisms are not the same as head turns. Prisms yield higher acuity because there is less fixation attempt. Fresnel prisms are not good substitutes for properly ground optical plastic. Many CN patients have both convergence and gaze angle nulls, and the former usually results in greater CN damping. This is important to know when the gaze angle null is large. Here, base-out prisms (with -1.0 spheres OU) can be used to converge the eyes, and the resulting damping will preclude a head turn.

Feedback of eye motion from inside the eyelids also may damp CN (24). This effect can be taken advantage of by prescribing soft contact lenses. Contact lenses with an anesthetic (to block afferent input) does not damp the CN. There is a simple procedure that may show if contact lenses will damp a patient's CN. Take a cotton swab and just touch the eyelid to see if the nystagmus damps; do not press on the eyelid. This method is an indicator of whether soft lenses might be useful to damp CN. Contact lenses should also be beneficial to patients without a null. Even if the cotton swab does not produce damping (perhaps owing to anxiety in the patient), it is easy to try contact lenses. Unfortunately, for

many years, contact lenses have not been prescribed for patients with nystagmus. Soft contact lenses at least (and possibly hard lenses) are not contraindicated by CN and may be indicated as a good therapy.

Surgical Treatment

Large-angle nulls are best treated surgically (20,25). Kestenbaum (26) and Anderson (27) independently described the recess/resect operation. This effectively rotates both eyes opposite to the null angle, such that the same innervation that preoperatively put the eyes at the null angle postoperatively puts them in primary position, and, therefore, the nystagmus is nulled.

If the patient not only has a null angle but also has strabismus and surgery is desired to minimize the nystagmus and straighten the eyes, the best approach is to operate on the good eye first, the fixating eye, not the strabismic eye. Any remaining tropia can be corrected after the initial operation. Sometimes, the surgery performed on the fixating eye reduces or eliminates the tropia. Sometimes, it is made worse. Moving the tropic eye so it is straight will not affect the nystagmus. If the strabismic eye was esotropic and the null was in adduction of the fixating eye, the surgical rotation has to be abduction. That will result in less esotropia and is the most common case. Esotropia is more common than exotropia, and the combination of esotropia and a null in adduction is more common than esotropia and a null in abduction. This causes a good surgical result on the tropic eye without surgery of that eye.

One should never use head turn to either indicate the amount of surgery or assess the results of surgery because head turns are under the control of the patient. It is best to measure the null using objective eye movement recordings, both before and after surgery, by having the patient fixate at different gaze angles with the head fixed so that there is no input from the patient (20). Evaluated that way, whatever null is measured on day 1 after the operation will be measured 5 years later. It will not have moved because it was fixed by the surgery (25). The surgery itself broadens the null, and lateral to the null, the nystagmus is less than before surgery. It appears to be a self-prophecy that any surgical rotation on a CN patient with an eccentric null and no convergence null produces a better result than not doing the surgery. Therefore, surgery moves the null angle, increases the null area, and decreases off-null nystagmus, and the shifted null is stationary. If surgery does not totally shift the null to primary position and the shifted null is a few degrees to one side, another surgery is not always necessary. Prisms could be used to shift the visual world to the postoperative null position and fine-tune the surgery.

Another surgical approach is a bimedial recession for patients who use convergence to damp the nystagmus. This is artificial divergence created by weakening either one or both medial recti (28). It might be contraindicated if the patient has strabismus, since it depends on the ability to fuse to straighten the eyes. Weakening both medial recti creates an artificial divergence, and the convergence innervation required to fuse an image nulls the nystagmus. It should also be considered when there are both convergence and gaze angle nulls, since the convergence null probably will yield the greatest CN damping. After a bimedial recession, the patient's distance refraction may need additional -1.0 spheres OU to compensate for the induced accommodation (similar to the use of base-out prisms).

The Faden operation times four weakens all four horizontal muscles (by Cüppers) (29,30). A disadvantage may be that double vision occurs in lateral gaze. Weakening the muscles reduces the effect of all innervation. Whether the nystagmus, or anything else, is going to be affected so that the nys-

tagmus will damp in primary position (or in all positions) requires the muscles to have less efficiency. This operation is useful in patients without nulls.

A minority of CN patients adopt a vertical or torsional head position to damp their nystagmus. Such patients can be treated with either prisms or surgery on the vertical or torsional extraocular muscles. Those with both CN and LMLN may be helped by the methods above if they exhibit primarily CN and by those below if they exhibit primarily LMLN. The key to successful therapy in these cases is in accurate analysis of the waveform characteristics and compensation mechanisms used to damp the nystagmus.

Other Treatment

Biofeedback has been shown to be useful in damping CN with repeated training (31, 32). Biofeedback is ineffective under conditions of stress when better acuity is needed.

Drugs are not beneficial in the treatment of CN. Despite reports of some CN damping with barbiturates or baclofen, the secondary effects of these drugs preclude their recommendation.

Injection of botulinum toxin into the extraocular muscles, has been claimed to damp CN, but the effectiveness of this procedure has not been studied adequately, and it must be repeated every few months to remain effective. Acupuncture is also being studied as a possible treatment for CN (33).

Finally, we have discovered that stimulation of the ophthalmic division of the trigeminal nerve may damp CN (34). This response, when present, is robust and stimulus independent. Touch, pressure, vibration, and subliminal electrical stimulation all have been found to damp CN. Making the transition from experimental to therapeutically useful methods of stimulation remains to be done.

LATENT/MANIFEST LATENT NYSTAGMUS

Differential Diagnosis

Manifest latent nystagmus, as first defined by Kestenbaum, is present with both eyes open but only one being used for fixation (35,36). There is always a tropia, and vision from the tropic eye is suppressed in the cortex (37). The slow phase is a decreasing velocity exponential, and the corrective fast phase is always in the direction of the eye that is fixating, the straight eye. MLN is the same nystagmus as the rare LN that only appears with occlusion of one eye. In MLN, the subject occludes one eye by cortically suppressing it while fixating with the other eye. Thus, LMLN refers to this single type of nystagmus that is present in most patients with both eyes open while one is fixating. In some patients, however, it may only be present when one eye is occluded.

The seldom seen condition of individuals with both CN and LMLN is difficult to diagnose correctly (3). One or the other might be dominant and result in complex waveforms and variations of nystagmus type with gaze angle. The best approach to diagnosing these patients is to use the methods required to accurately diagnose the more straightforward types.

Diagnostic Criteria

Waveform

Waveform is also the diagnostic criterion for LMLN (Table 2). Recordings show a decreasing velocity exponential slow phase (Fig. 1). All patients (100%) with LN or MLN have strabismus. This definition of strabismus includes latent strabismus (i.e., the phoria resulting when one covers an eye). Thus, LMLN includes pure LN, where the eyes are straight with both eyes

open, and when one eye is covered, it will develop an esophoria or exophoria followed by LN in both eyes. MLN mimics LN exactly if it is bidirectional. When MLN is unidirectional and the patient fixates with one eye, there will be no nystagmus and the other eye will be esotropic, but when the patient fixates with the other eye and the formerly fixating eye is esotropic, he or she will have MLN. Pure LN (i.e., no nystagmus with both eyes open) is rare. If you occlude the left eye and the right eye is fixating, jerk right LN with decreasing velocity slow phases results, and vice versa. LN implies strabismus, but the reverse is not true. Distinguishing the LMLN waveform and the tropia of the nonfixating eye requires DC-coupled, high-bandwidth recordings of both eyes simultaneously.

The small group of patients with both CN and LMLN present a diagnostic nightmare. Some have mostly CN (CN/LMLN), and their waveforms are any of the CN waveforms (i.e., pendular or increasing velocity slow phases) and one other waveform called *dual jerk latent* (Table 2). The latter is a waveform in which pendular is superimposed on a decreasing velocity slow phase jerk waveform. They do not exhibit the pure latent waveform (i.e., decreasing velocity slow phases). Therefore, CN is predominant. The other group has mostly LMLN (LMLN/CN), and their waveforms are latent and dual jerk latent. There are some who have equally CN and LMLN. At various times, they exhibit the CN waveform, latent waveform, or the dual jerk waveform. A linear slow phase is not diagnostic of either CN or LMLN. When a pendular waveform is superimposed on a jerk waveform and the slow phase is accelerating, it is a congenital dual jerk. If the slow phase is decelerating, it is a dual jerk latent. One has to determine carefully what is happening to the axis of the pendular slow phase (i.e., whether it is decelerating or accelerating) to properly categorize the nystagmus. This small but difficult group of patients must be recorded for accurate diagnosis and treatment, since treatment depends on which component (CN or LMLN) is dominant. Distinguishing the CN and LMLN waveforms from the combination waveforms requires DC-coupled, high-bandwidth recordings of both eyes simultaneously.

To summarize, within the different types of benign infantile nystagmus, there is a large category of pure CN, a significant category of pure LMLN, and a small category that is a mixture of the two. There also are individuals with spasmus nutans or NBS. All are easily diagnosed with the aid of ocular motility recordings and just as easily misdiagnosed without them. There are 12 CN waveforms, 1 latent waveform and 1 mixed waveform (dual jerk latent). A large percentage of patients (80%) will have CN and 15% LMLN, with only a small percentage having mixtures of both. If you consider only CN patients, 94% will have pure CN and 6% a mixture. If you restrict the population to LMLN patients, 75% of them will have only LMLN, and the rest will have mixtures of CN and LMLN. Thus, more patients with predominantly LMLN will also have some CN than patients with predominantly CN having LMLN.

Symptomology

As stated previously, pure LN is extremely rare (31,38). That is, when you record pure LN, you must find no nystagmus with eyes open at all gaze angles. There have been only a few cases proven by recordings to have pure LN. Many patients who clinically appear to have pure LN actually have MLN. When one records these patients, the MLN is visible, and small LN can be seen with an ophthalmoscope. Pure LN in primary position with MLN in lateral gaze is more common, and most common is MLN at all gaze angles.

The intensity of LMLN is greatest with gaze toward the direction of the fast phase (Alexander's law). Jerk right nystagmus is

greater in right than in left gaze, and vice versa. These are not true nulls. One cannot show increased nystagmus because the eye is at the end of its excursion. Instead, this is an example of a monotonic relationship of gaze and amplitude. It would not be uncommon for a person who fixates with one eye and has MLN to keep that eye in adduction where Alexander's law will reduce the nystagmus. In CN there is a true null, demonstrating increased amplitude (with increasing velocity exponentials) as gaze is directed away from the null in both directions, whereas in LMLN (right eye or left eye fixating), there is an Alexander's law relationship and decreasing velocity exponentials. Patients with LMLN usually place their fixating eye in adduction to minimize the nystagmus and thereby maximize acuity. Some might also place the fixating eye in abduction due to a non-Alexander's law (inverted) variation. As in CN, the head turn minimizes the nystagmus and maximizes acuity. A patient might place his eye in other than the minimum position of nystagmus if he had an angle kappa that required eccentric fixation. Better acuity results, although the nystagmus might be a little higher where the patient places gaze.

We have never recorded the LN waveform in patients with orthophoria. They all had latent strabismus (when you cover one eye, the other does not remain straight). MLN has never been recorded in patients with binocular alignment. All had manifest strabismus. Both eyes are open, but one eye must be deviated (in or out) to have MLN. If they can straighten their eyes, the MLN disappears. In the blockage syndrome, they have CN when their eyes are aligned. Therefore, LN and MLN imply strabismus. Strabismus does not imply LMLN (50% have no nystagmus at all), but if an individual has LN, he also has strabismus. Even if it is not evident clinically, a microstrabismus can be recorded. Strabismus is a necessary but not sufficient condition for LMLN. It is either a latent strabismus, in those rare patients with pure LN, or a tro-

pia for MLN. Summarizing, CN can occur with or without strabismus, but all MLN patients have strabismus.

Treatment

Surgical Treatment

It is possible to surgically enhance the acuity of some patients with LMLN (39). If a patient has LMLN and a constant head turn to take advantage of an Alexander's law reduction of the nystagmus, he is fixating with his adducted eye (the other being in a tropic position and suppressed). Under these conditions, one may operate on the fixating eye to move the low-amplitude LMLN from the eccentric gaze position to primary, that is, rotate the fixating eye to an abducted position so the innervation that previously placed it in adduction will result in gaze straight ahead. If the strabismus accompanying the LMLN was esotropia (the most common case), the strabismic eye will assume a less tropic position after rotation of the fixating eye because of the innervation required to straighten the latter. This procedure may correct the strabismus in addition to markedly reducing the nystagmus in primary position. Any remaining strabismus can be corrected later by rotating the strabismic eye. If the strabismus is exotropia, this procedure will tend to worsen it, but a second operation on the strabismic eye can correct this condition. If the LMLN patient exhibits two head turns and alternately fixes with the adducted eye, this procedure should not be used. Here, a Faden operation may prove useful.

SPASMUS NUTANS

Differential Diagnosis

Spasmus nutans may appear at or after birth and usually, but not necessarily, ceases by the age of 3. The nystagmus is pendular, usually monocular or disconju-

gate, and commonly accompanied by a compensatory head oscillation. Reports of "spasmus nutans" accompanying neurologic disease have not included recordings to prove that the nystagmus was that of spasmus nutans.

Diagnostic Criteria

Waveform

The diagnostic criteria for spasmus nutans have been defined by ocular motility recordings (40). We can now diagnose spasmus nutans immediately. It is no longer necessary to wait 3 or 4 years before making the diagnosis based on its possible disappearance. The waveform is a dissociated pendular nystagmus, and this dissociation may be so great that the nystagmus is uniocular (Table 2). The diagnostic key is the variable phase difference between the oscillations in both eyes, unlike CN and LMLN where the oscillations are conjugate. The spasmus nutans waveform can vary anywhere from pure conjugacy to pure disconjugacy, 0 to 180 degrees phase shift. It varies during the recording, usually from minute to minute. Distinguishing the variable phase relationship between the pendular oscillations of both eyes requires DC-coupled, high-bandwidth recordings of both eyes simultaneously.

Symptomology

The head nodding in spasmus nutans, unlike CN, is compensatory (41). The ocular oscillations are asymmetric between the two eyes, and contrary to common belief, it does not always disappear. Patients of 10 or 12 years of age have been reported to have spasmus nutans. Many times, it disappears to clinical observation (as MLN), but when recorded, a pendular dissociated nystagmus can be demonstrated. The head nodding in spasmus nutans is curious. The VOR of these patients is normal, but by shaking the head willfully, the nystagmus is switched off and the eyes become stable in space (due to normal VOR). As a result, their acuity increases. A patient may have convergence nystagmus, one eye going left, the other eye going right at the same time (180 degrees out of phase), while the head is still. When the head starts shaking, the nystagmus stops. Owing to a normal VOR, the eyes begin moving conjugately equally and opposite to the head, and gaze remains constant. Thus, patients cancel the disconjugate pendular oscillation of spasmus nutans (present with a still head) and substitute a conjugate VOR when the head is moving, and acuity increases. The oscillating diplopia that probably results from the out-of-phase oscillations may be the main reason that head-shaking is used to cancel the nystagmus.

Treatment

Spasmus nutans is benign and requires no treatment.

NYSTAGMUS BLOCKAGE SYNDROME

Differential Diagnosis

The NBS is the source of some misunderstanding. Individuals with NBS use a convergence-like movement to damp their CN while viewing a distant target. Thus, the waveforms in NBS are those of CN when the patient is looking in the distance and the eyes are straight (42). With the imposition of the purposive esotropia (this is not a strabismus that occurs transiently but one that the patient willfully imposes because he has found that acuity is better under that condition), the waveform can either become a damped CN waveform (one type of NBS) or a small-amplitude MLN (another type of NBS). Thus, there are two types of blockage syndrome, both exhibiting CN waveforms before the imposition of esotropia. NBS is often misdi-

agnosed in patients with LMLN and alternating fixation who place their fixating eye in adduction to reduce the LMLN. Since they do not have CN, they do not have NBS.

Diagnostic Criteria

Waveform and Purposive Esotropia

The NBS criteria, as stated previously, are twofold: in one type, a CN waveform changes to a damped CN waveform, so acuity is better (that is why the patients induce the esotropia); in the other type, a CN waveform becomes the LMLN waveform, and even though that is usually a waveform unsuitable for good acuity (because there are no long foveation periods), it is of low amplitude and acuity increases. Obviously, to accomplish this, patients must have a variable strabismus. The recordings show one eye turned in without diplopia, and there is a variable, purposive strabismus. When fixating in the distance, the eyes are orthotropic. They become esotropic by design, and then either one (CN) or the other (MLN) low-amplitude waveform results, along with better acuity. Distinguishing the CN from the LMLN waveforms and the purposive esotropia of the nonfixating eye requires DC-coupled, high-bandwidth recordings of both eyes simultaneously.

Symptomology

Patients with NBS willfully induce esotropia while fixating at distance. This is something that most with CN cannot do unless they have the ability to partially suppress the esotropic eye. If CN patients with normal binocularity could make one eye esotropic, they would have oscillating diplopia. When NBS patients turn one eye in and suppress it, their nystagmus damps, and they may adopt a head turn to place their fixating eye in adduction. The CN is either reduced in amplitude or converted to a low-amplitude LMLN by this purposive esotropia. The greater the esotropia, the lower the nystagmus, and as the fixating eye moves from adduction to abduction, the nystagmus increases and esotropia decreases.

Treatment

Surgical Treatment

In the blockage syndrome, a bimedial recession might prove beneficial, since the purposive strabismus is different from ordinary strabismus. That bimedial recession may help those with NBS has been suggested, but published studies are lacking. Similarly, the Faden operation may benefit those with NBS. Simple surgical correction in NBS has not proven successful (43).

CONCLUSIONS

It is now accepted that electrophysiologic tests (ERG and VEP) are needed to assess afferent visual system function at both the retinal and central levels. Unfortunately for the patient with nystagmus, the use of accurate ocular motility recording for the dual roles of diagnosis and treatment is not widely used. Only in selected centers of ophthalmologic or neuroophthalmologic research are such motility recordings routinely made. Years ago, cardiologists realized that they could not rely solely on their ears to accurately diagnose complex disorders of cardiac function. Too many ophthalmologists are still attempting to use only their eyes for the same purpose. It simply cannot be done reliably for ocular motor disorders. To paraphrase the magician, "the (patient's) eye is quicker than the (physician's) eye."

In this chapter, I have summarized the results of the past 20 years of ocular motor research relevant to the diagnosis and treatment of CN and other benign types of nystagmus of infancy. Analogous to the use of

ERG and VEP, definitive diagnostic motility criteria have been established for all types of infantile nystagmus. Clinical signs of the various types of nystagmus often are ambiguous, and reliance on them without quantitative motility data can cause diagnostic errors that become compounded by either wrong choices for therapeutic intervention or inaccurate estimations of null angles on which surgery is based. In many cases, visual acuity can be improved or binocularity can be restored by the correct therapy applied in a timely manner. The patient's complete diagnosis must include the motility diagnosis in addition to the afferent system diagnosis, as must the therapeutic plan. This requires the addition of routine eye movement recordings in the clinical setting to augment ERG and VEP.

ACKNOWLEDGMENTS

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The Vestibulo-Ocular Reflex and Vertigo

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Raven Press   New York