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Management of Congenital Nystagmus

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Congenital nystagmus (CN), as the term is used herein, is a specific type of nystagmus that usually presents in early infancy; there are other types of nystagmus of infancy that are *not* CN. The word *congenital* means "with birth," and although CN may be present at birth, it may also become manifest later in infancy, childhood, or even adulthood. Thus, this term should not be taken literally but rather 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. It is pointless to attempt to change the name of this motor sign to one that merely shifts the time of onset (such as "infantile" nystagmus), since this 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 chosen name should reflect the waveform(s) that are intimately related to the underlying mechanism(s) rather than some arbitrary time of onset; the latter only serves to exclude those with the *same* disorder but different timing. To paraphrase Marshall M. Parks (discussing "congenital" versus "infantile" esotropia), "To adopt another term in place of the widely used congenital *nystagmus*, just because the *nystagmus* cannot be proven to be present at birth may not be in the best interest of *nystagmography*" (italics added to indicate changed words). The most important factor in the management of CN is not its name but its accurate diagnosis and differentiation from other benign types of nystagmus of infancy and from symptomatic acquired types.

I. DEFINITION AND DIFFERENTIAL DIAGNOSIS

CN is a motor eye sign and 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 visual target and back to the target (*not* across the target). The pendular waveforms look sinusoidal but they are usually distorted by both flattening and presence of small foveating saccades on the peaks corresponding to where the foveation occurs. The jerk waveforms 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 that runaway. This saccade might return the eyes back to the target ("foveating" saccade) or initiate a slow eye movement back to the target for refoveation. The direction of nystagmus is defined by the direction of that saccade despite the fact that it is the slow eye movement that causes the nystagmus. CN is usually horizontal but occasionally may have vertical components in some patients; torsional components are not uncommon.

Also common are associated sensory defects (Table 1) that may accompany CN of all waveforms. One cannot equate pendular CN with a sensory defect or jerk CN with a motor defect; *all* CN waveforms are due to a primary motor defect. We have determined that the etiology and the mechanisms of these nystagmus waveforms are independent of accompanying sensory defects and therefore, although there is an association between many sensory defects and the presence of CN, it is not a causal relationship. The sensory defect is neither the primary cause of the nystagmus nor is the nystagmus the primary cause of the defect. Sensory defects appear to be secondary factors provoking the manifestation of CN in individuals in whom there is already a primary predisposition for the latter. Thus, although sensory defects are not the primary cause of CN, they appear to be the precipitating factors in the manifestation of CN in many patients. The observation, that *any* of several different sensory abnormalities can result in the developing motor system becoming unstable, leads to the following possibilities: (1) a small percentage of individuals are born with a motor system

Table 1 Sensory Defects Associated with Nystagmus

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

that is precariously close to oscillation, or (2) nature has evolved the ocular motor system in such a way that the horizontal system is close to instability in many individuals. The reason for the latter hypothesis may lie in our need for rapid horizontal eye motion to survive. Whatever the underlying reason, the motor system seems to require early visual input to assure its stability and any sensory defect interfering with vision might result in the instability we call CN. In those individuals actually born with CN, the system may have developed with such a strong instability (due to hereditary or spontaneous genetic factors) that even the presence of adequate visual input after birth is not enough to stabilize it.

When there is a sensory defect, it rather than the nystagmus may be the limiting factor for acuity, and even halting eye motion using forceps may not result in an acuity improvement in some patients; in others, acuity can be improved by damping the CN. One must perform the necessary diagnostic tests (e.g., the electroretinogram [ERG]) to assess correctly the functional integrity within the visual system and diagnose any sensory abnormalities present, but that diagnosis is *not* sufficient to describe the motor defect causing the nystagmus. The latter requires motility recordings for absolute diagnosis. If a patient with nystagmus has a visual acuity uncorrectable to better than 20/60, one should look carefully for afferent defects. Sensory defects may be more prominent in pediatric patient populations than in adult populations where there is a higher incidence of idiopathic CN. It is possible that there are two different genes with variable probabilities of the one causing the slow eye movement instability CN and the other causing the afferent defect. The presence of either of these genes might be related to the presence of the other by some third probability. Another possibility is that there is one gene with different probabilities for an afferent defect and CN; a patient may have both or one or the other. The diagnosis and treatment of sensory abnormalities will not be covered in this chapter that concentrates on the proper management of CN and other forms of infantile nystagmus.

In addition to CN, other types of nystagmus (and saccadic oscillations) may also occur at or shortly after birth and should not be confused with, or lumped with, CN. They are different from CN in waveform (mechanism) and clinical characteristics. Some of these other benign types of infantile nystagmus are: latent/manifest latent nystagmus (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 (1) determining whether this is a benign nystagmus or one that is indicative of disease, and (2) if the nystagmus is benign, determining whether it is CN, LMLN, spasmus nutans, or NBS. Those types of nystagmus that have been determined to be symptomatic types are downbeat nystagmus (due to structural brain stem abnormalities), epileptic nystagmus, uniocular nystagmus (due to optic nerve glioma), vestibular nystagmus (due to any number of vestibular problems), and "nystagmus" of the blind; the latter is not truly a nystagmus but rather wandering

eye movements. Table 2 lists both benign and symptomatic forms of nystagmus of infancy. Another instance when it is important to distinguish between CN or a benign or symptomatic nystagmus is 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 unclear whether the nystagmus was present before the trauma or especially if it occurred following the trauma.

Manifest latent nystagmus (MLN) is present with both eyes open but only one being used for fixation. There is always a tropia, and vision from the tropic eye is suppressed in the cortex. The slow phase is a decreasing-velocity exponential, and the corrective fast phase is always in the direction of the eye that is fixating; that is, the straight eye. MLN is the same nystagmus as the rare, latent nystagmus 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, the term *latent/manifest latent nystagmus* refers to this type of nystagmus that is present in most patients with both eyes open while one is fixating but, in some patients, may only be present when one eye is occluded.

Spasmus nutans may appear at or after birth. It is pendular and usually, but not necessarily, ceases by the age of 3 years. It also is usually monocular or disconjugate and is commonly accompanied by a head oscillation. Reports of "spasmus nutans" accompanying neurological disease have not included recordings to document that the nystagmus was that of spasmus nutans.

The nystagmus blockage syndrome is the source of some misunderstanding. NBS waveforms are those of CN when the patient is looking in the distance and the eyes are straight. When there is a purposive esotropia (this is *not* a strabismus that occurs transiently but one that the patient willfully imposes because he or she 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. NBS

Table 2 Types of Nystagmus of Infancy

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

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

is often misdiagnosed in patients with LMLN and alternating fixation who place their fixating eye in adduction.

Unfortunately, for the diagnostician, there exist individuals with both CN and LMLN (it is not usual, but some patients have a combination of CN and LMLN); one or the other might be dominant and result in complex waveforms and variations of nystagmus type with gaze angle. These become very difficult to diagnose and the best approach is first to learn to diagnose accurately the more straightforward types.

II. DIAGNOSTIC CRITERIA

Unambiguous, diagnostic characteristics exist for each of the benign types of nystagmus of infancy. Simple eye movement recordings differentiate these types and simultaneously provide the data necessary for effective therapeutic intervention.

A. Waveform

The waveforms recorded from infants with CN where the nystagmus was noted at birth are the same as those seen in infants where they were noted to appear later. Since the waveforms are the same, the mechanism in both instances is the same; 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 3) that have been identified in CN are pathognomonic of CN except pure pendular; that could be an acquired nystagmus. Usually the pendular nystagmus of CN is distorted so the patient can foveate, whereas in acquired nystagmus it is not. In acquired nystagmus, the slow phases can be linear, of increasing velocity, of decreasing velocity, or pendular. CN slow phases can be pendular, increasing velocity, or some may look linear. Within the pendular or jerk major types of CN, we identified three different forms of pendular and eight jerk, four unidirectional and four bidirectional. One waveform, dual jerk, is a superimposition of pendular on a jerk waveform with increasing velocity slow phases; illustrations and examples are available in the literature. In a group of patients with CN, 87% had more than one waveform; they had any number of the 12 waveforms. A small percentage of patients (13%) exhibited only one waveform.

One specific group of patients who usually have CN are those with albinism. Here, as with CN, what was once a "clinical" diagnosis can now be made definitively by a simple diagnostic test based on the visual evoked potential (VEP). If performed properly using the paradigm developed by Apkarian, the VEP is virtually 100% free of both false positives and false negatives. This paradigm uses full-field, monocular pattern (checkerboard) onset/offset stimuli

Table 3 Nystagmus Waveforms

Congenital	Latent
Pendular	
Pendular	
Asymmetric pendular	
Pendular with right/left foveating saccades	
Unidirectional jerk	
Jerk right/left	Jerk right/left
Jerk right/left with extended foveation	
Right/left pseudocycloid	
Pseudo jerk right/left	
Bidirectional jerk	
Pseudopendular	
Pseudopendular with right/left foveating saccades	
Triangular	
Bidirectional jerk right/left	
Dual	
Dual jerk right/left	Dual jerk right/left latent

rather than 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. Appropriate VEP testing demonstrates the unequivocal dissociation between the misrouted fibers found in albinos and their CN, since no individuals with hereditary or idiopathic CN had misrouting. Thus, the hypothesis that misrouting can cause CN lacks scientific foundation.

The diagnostic criterion for LMLN also is the waveform (see Table 3); recordings reveal a decreasing velocity exponential slow phase. All patients (100%) with latent (LN) or manifest latent nystagmus (MLN) have strabismus. I include in the definition of strabismus the latent strabismus (phoria) resulting when you cover an eye. Thus, there is pure LN, where the eyes are straight with both eyes open and when you cover one eye, it will develop an esophoria or exophoria followed by LN. The LN waveform has decreasing velocity slow phases. Pure latent nystagmus (i.e., no nystagmus with both eyes open) is very 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. Latent nystagmus implies strabismus but the reverse is not true.

The diagnostic criteria for spasmus nutans have also been defined. We can now diagnose spasmus nutans immediately by recording; 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. The diagnostic key is the variable phase difference between the oscillations in both eyes; unlike CN and LMLN that are conjugate. The spasmus nutans waveform can vary anywhere from pure conjugacy to pure disconjugacy; 0- to 180-degree phase shift. It varies during the recording, usually from minute to minute.

The nystagmus blockage syndrome criteria, as indicated above, are twofold: In one type, CN changes to damped CN, so acuity is better (i.e., why the patients induce the esotropia); in the other type, the CN becomes LMLN 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. If one is binocular, one cannot turn one eye in without diplopia resulting. These patients turn one eye in and they do not experience diplopia (they suppress that eye); they have a variable, purposive strabismus. When they are looking in the distance they are orthotropic; they become esotropic by design and then they either exhibit one (CN) or the other (MLN) low-amplitude waveform along with better acuity.

There is a very small group of patients who present a diagnostic nightmare—CN and LMLN together. Some of them 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" (see Table 3). The latter is a waveform where pendular is superimposed on a decreasing velocity slow phase jerk waveform. They do not have pure latent waveforms (i.e., decreasing velocity slow phases) so therefore CN is predominant. The other group has LMLN/CN, and their waveforms are latent and dual jerk latent. There are some patients 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. If the slow phase is accelerating, it is a congenital dual jerk. If the slow phase is decelerating and a pendular waveform is superimposed, it is a dual jerk latent. One has to determine carefully what is happening to the axis of the pendular slow phase (is it decelerating or accelerating?) to categorize properly the nystagmus. This small but difficult group of patients must be recorded for accurate diagnosis.

Summarizing, 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; all are easily diagnosed with the aid of recordings. There are 12 CN waveforms, one latent waveform and one mixed waveform (dual jerk latent). If a patient walks into your office with wiggling eyes and you wish to guess what they have before you record them, your best guess would be CN. A large percentage (80%) will be CN and 15% LMLN with only a small percentage being mixtures. If you look at just patients with CN, 94% will be pure CN and only 6% a mixture. If you

just look at patients with LMLN, three quarters of them will just have LMLN, but there is a significant number of patients who will have mixtures. Thus, more patients with predominantly LMLN will also have some CN than patients with predominantly CN have LMLN.

III. CLINICAL CHARACTERISTICS

A. Congenital Nystagmus

In addition to the quantifiable and diagnostic characteristics of CN available through eye movement recordings, there are clinical signs one can look for in the office. In CN, one should look for a null angle; an indication of this 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 vision because it is not "appropriate" to walk around with their head turned, but a child will have a head turn. A positive family history and negative neurological examination are indicative of CN.

If patients hold reading material close, one should suspect that converging the eyes will damp the nystagmus. They may have a latent component; this can be checked by the cover test. If the nystagmus direction reverses, one still does not know whether it is LMLN or CN with a latent component (they *are* different). Such patients may also have head nodding that is not compensatory.

The CN magnitude damps at a specific gaze angle if there is a null. The definition of a null (a true null) is that the nystagmus must get larger on either side. This is in contrast to LMLN where monotonic variation with gaze angle (Alexander's law) causes patients to keep their eyes deviated to one side where the nystagmus is low. This is not a true null, because there is no increase on both sides. 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. The null angle during fixation does not usually equal the null angle during pursuit, optokinetic nystagmus, or head movement (where the vestibulo-ocular reflex is stimulated). Usually, the null is shifted in the direction opposite to the eye movement. During pursuit to the left, the null moves to the right of the static null; and during pursuit to the right, the null moves to the left. In the CN population, 48% have both convergence and gaze angle nulls, 29% only convergence nulls, 9% only gaze angle nulls, and there are 14% with no nulls.

Children with CN automatically adopt a head turn to see better. They do not have to wait to be told about the null, they know where it is because things appear clearer when they turn their head. One may think of the null as a region of ocular motor equilibrium. The brainstem (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, the acuity may not increase measurably.

Sometimes the patient will tilt the head and sometimes turn the head (vertically or horizontally) and tilt it. Perhaps the oblique muscles are involved; this has not really been studied well. It represents innervation of muscles other than the horizontal muscles that are somehow helping to reach equilibrium in what are basically horizontal oscillations. Most CN is horizontal with little or no vertical components. Few patients have significant vertical components, and many may have small torsional, components. Again, acuity increases with vertical or torsional head positions, especially if there are no afferent defects.

The CN might reverse direction on covering an eye, because (similar to when the person pursues) the null moves, and therefore you may see a direction reversal of the CN if gaze went from one side of the null to the other. MLN mimics latent nystagmus 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. Although CN with a latent component looks like LN or MLN, it is not because the waveform remains CN; only the CN direction changes, because the null has moved with occlusion.

Many children with CN exhibit spontaneous head oscillations. When adults with CN concentrate on a visual task (real or imaginary), they may also have spontaneous head oscillations, but they usually learn to keep the head still because it is socially unacceptable to allow the head to shake. These head oscillations use existing pathways in the neck muscles. EMG in the neck muscles show that when normal individuals 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 the 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 accomplish anything; it is a manifestation of an existing oscillation on existing pathways to the neck. It used to be 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 very 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. When you understand the VOR, you know this compensatory hypothesis cannot work. For realistic compensation, 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 stability only during that part of the waveform and is a possible form of compensation useful only when the foveation periods do not result in stable eyes with the head still.

In patients with CN, the VOR is normal; it is not affected by CN. The head oscillations are merely an extension of the CN, and during the foveation periods, the eyes do not move and acuity is unaffected by head movements. The head oscillations in CN with a normal VOR are equivalent to those of a normal person moving the head and acuity does not change. In a few patients, if the VOR is suppressed and the head moves opposite to the eyes only during the foveation periods, these periods become motionless in space, because they were not so before the head was moving; here acuity can increase. Thus, if the CN waveform does not contain good foveation periods, the head might oscillate in a compensatory way so that during the foveation period, gaze remains stable in space (i.e., the eye movement recording is flat). If patients have good foveation periods, there is no reason to shake the head. Thus, if the foveation periods are flat, head motion cannot help the patient.

B. Strabismus

What is the relation of strabismus to nystagmus (to the CN or the LMLN)? The presence of LMLN means that there is strabismus. Of all patients with strabismus, 50% do not have any nystagmus. The other 50% do have nystagmus of some type; the question is, what type? Of that 50% of the total, 26.5% will have CN, only 17% will have LMLN, and 6% will be mixed. If one considers 100 patients who walk into the office with both nystagmus and strabismus and one has to guess (even though all patients with manifest latent nystagmus must have strabismus), the best guess is CN, because there are many more patients with CN than with LMLN. For 100 patients with both CN and LMLN, 43% had strabismus and 57% had no strabismus. So, the patient who has nystagmus probably does not have strabismus; more than half will not. If one considers just patients with CN (including those with some LMLN), almost three quarters do not have strabismus; thus, most patients with CN do not have strabismus. If one considers only those with pure CN, the great majority of patients do not have strabismus. We have recorded all 12 CN waveforms in patients with binocular alignment, so it is not necessary to have strabismus for any particular CN waveform. Of 43% of patients with strabismus in the first 100 mentioned above, more than half had CN even though most patients with CN do not have strabismus. Summarizing, for strabismus with nystagmus: 53%—CN, 35%—LMLN, and 12%—mixed.

C. Latent/Manifest Latent Nystagmus

Pure latent nystagmus is extremely rare. That is, when you record pure LN, you will find no nystagmus with eyes open at all gaze angles. There have been only

a couple of cases proven by recordings to have pure LN. Many patients thought clinically to have pure LN actually have MLN. When one records them or examines them with an ophthalmoscope, the MLN is visible. More common is pure LN in primary position with MLN in lateral gaze; 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 patient is at the end of the excursion of the eye; 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 to have MLN with that eye in adduction where Alexander's law will reduce the nystagmus. In CN, there is a null and 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 LN usually place their fixating eye in adduction to minimize the nystagmus and thereby maximize acuity. Some might also place the fixating eye in abduction because of a perverted non–Alexander's law variation. As in CN, the head turn minimizes the nystagmus and maximizes acuity. A patient could place the eye in other than the minimum position of nystagmus if he or she had an "angle kappa" that required eccentric fixation. Better acuity results despite the fact that the nystagmus might be a little higher where the patient places the 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 turned in or out to have MLN. If patients can straighten their eyes, the MLN disappears; in the blockage syndrome, patients have CN. Therefore, LN and MLN imply strabismus. Of course, strabismus does not imply LMLN (50% have no nystagmus at all), but if an individual has LN, strabismus also is present. Even if it is not evident clinically (it may be a microstrabismus that can be recorded), it is there. 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 tropia for MLN. Thus, CN can occur with or without strabismus; all patients with MLN have strabismus.

D. Spasmus Nutans

In spasmus nutans, unlike CN, the head nodding is compensatory. The ocular oscillations are asymmetrical between the two eyes and, contrary to what is commonly believed, it does not always disappear. There have been 10- or 12-year-old patients who still had spasmus nutans. Many times spasmus nutans

disappears to clinical observation (again, like MLN), but when recorded, a pendular dissociated nystagmus will be found. The head nodding in spasmus nutans is very 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 because of a good VOR; their acuity increases. A patient may have convergence nystagmus, with one eye going left and 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 oppositely to the head, so 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.

E. Nystagmus Blockage Syndrome

Patients with NBS can willfully become esotropic at distance. This is something a person with CN cannot do unless they have the ability partially to suppress the eye they purposely make esotropic. If patients with CN with normal binocularity made an eye esotropic they would have diplopia—oscillating diplopia. Patients with NBS can turn one of their eyes in and suppress it; their nystagmus damps and they adopt a head turn to place their viewing 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. These patients usually put the fixating eye in adduction.

IV. THERAPEUTIC OPTIONS

What are the therapeutic options for the various types of benign nystagmus of infancy? In CN, we have found that a strong fixation reflex maintains foveation despite slow acceleration off the target. This again is juxtaposed to the mistaken idea that CN is caused by poor fixation reflexes. We have found that fixation in CN is very accurate (<13 min of arc). In LMLN, the fixation reflexes seem to be ineffective, since the slow phase immediately takes the eyes away from the target as soon as a fast phase is completed. These patients seem to have no ability to use the fixation system to keep the eye on the target. One can increase the visual acuity of patients with CN (if they have no foveal defects) if there is a null angle or a convergence null and if there are good waveforms with long foveation periods. In such patients, prisms or surgery may be effective, but if a patient does not have these characteristics, these methods are not likely to be successful. One could still straighten the head, but these characteristics are

necessary for an increase in acuity. If one can minimize the attempt to fixate, acuity will be maximized, because in CN, the greater the effort to see, the higher the level of nystagmus. Even in turning the head to find the null, the level of nystagmus of that null is higher than if the null is placed straight ahead surgically. That null is better because there is no effort involved. Visual acuity is proportional to how much time in each cycle the target is foveated. 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/s. Data from normal individuals show that when retinal slip stays below 4 degrees/s, 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" in engineering terms. 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 of these characteristics must be present for good acuity—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. It can be used to plan surgery, optical correction, or other therapy.

A. Optical Therapy

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 substitutes for head turns; they yield higher acuity than head turns, because there is less fixation attempt. Fresnel prisms are not good substitutes for properly ground optical plastic. It should be noted that many patients with CN have both convergence and gaze-angle nulls and the former usually result in greater CN damping. This is important to know when the gaze-angle null is large; here base-out prisms (with –1.0 spheres) can be used to converge the eyes and the resulting damping will preclude a head turn.

Feedback of the eye motion from the inside of the eyelids may also damp CN. This effect can be taken advantage of by inserting soft contact lenses in a patient with CN. Contact lenses with an anesthetic (to block afferent input) do not damp the CN. There is a simple office procedure that may indicate 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. It is a quick way to

tell whether the soft lenses might work. Contact lenses should be beneficial to patients without a null. Even if the cotton swab does not produce damping (perhaps due 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 are *not* contraindicated by CN and may be indicated as a good therapy.

B. Surgery

Large-angle nulls are best treated surgically. Kestenbaum and Anderson independently developed the recess and 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 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 in a good surgical result on the tropic eye without surgery of that eye.

One should never use head turn to assess the results of surgery. The young patient realizes the parents are spending a lot of money and hears the physician say that the surgery is going to make his or her head straight; one cannot trust the patient's postoperative head turn on the first visit after the surgery because that patient is going to keep the head straight. This scenario is probably the cause of the myth that the surgery was correct (no head turn) and 6 months later the head turn came back. The head turn cannot be used to assess the surgical result because it is under control of the child who is subject to all kinds of input; the patient may want to please the parents or the surgeon. It is best to measure the null by having the patient fixate at different gaze angles with the head fixed both before and after the operation, so that there is no input from the patient. This is preferably done by recordings before the surgery to document the null and after the surgery to see where the null shifted. 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. The surgery itself, luckily for the surgeon, broadens the null (so even if the null was slightly miscalculated,

the results will not be off by much) and lateral to the null the nystagmus is much less than before surgery. It appears to be a self-fulfilling prophecy that any surgical rotation of a patient with CN with a null produces a better result than not doing the surgery; furthermore, the null is broader because the CN has damped at all gaze angles. Therefore, surgery moves the null angle, increases the null area, and decreases off-null nystagmus and the new null is stationary. If surgery does not totally move the null to primary position and there is a new null a few degrees to one side, another surgery is not always necessary. Prisms could shift the visual world to the new null and fine-tune the surgery.

Another surgical approach is a bimedial recession. This is artificial divergence created by weakening both medial recti. It might be contraindicated if the patient has strabismus, since it depends on the ability to fuse to straighten the eyes. Perhaps in the blockage syndrome it would still work because this is a different kind of strabismus—a purposive strabismus. Weakening both medial recti creates an artificial divergence and the convergence innervation required to turn the eyes to primary position nulls the nystagmus. This, of course, is in a person who has a convergence null; this is the condition under which this operation would be successful. It should also be considered when there are both convergence and gaze-angle nulls, since the convergence null will probably yield the greatest CN damping. Bimedial recession may also help those with NBS; this has been suggested but published studies are lacking.

The Faden operation times four weakens all four horizontal muscles (developed by Cüppers). Double vision in lateral gaze may be a problem with this operation, but some say it does not matter because the patient is happy. Weakening the muscles reduces the effect of all innervation. Whether the nystagmus, or anything else, is going to be affected so that the nystagmus 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 patients with CN adopt a vertical or torsional head position to damp the nystagmus. They 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 discussed above if they exhibit primarily CN and by those methods discussed below if they exhibit primarily LMLN. The key to these cases is in the accurate diagnosis.

It is also possible to enhance surgically the acuity of some patients with LMLN. If a patient has LMLN and a constant head turn to take advantage of an Alexander's law reduction of the nystagmus, he or she is fixating with the 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 a 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 owing to the innervation required to straighten the latter; this procedure may correct the strabismus in addition to 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 also correct this condition. If the patient with LMLN exhibits two head turns and alternately fixes with the adducted eye, this procedure should not be used. Here, a Faden operation may prove useful.

C. Other Therapies

Biofeedback has been shown to be useful in damping CN and it is claimed that with repeated training the effect can be used by the patient outside the laboratory without the biofeedback. This remains to be demonstrated under conditions of stress when higher acuity is needed for an important task.

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.

The injection of botulinum toxin into the extraocular muscles has also been reported to damp CN, but this procedure must be repeated every few months to remain effective.

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

V. CONCLUSIONS

The importance of electrophysiological tests (ERG and VEP) to assess afferent visual system function at both the retinal and central levels is now accepted. 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 ophthalmological or neuro-ophthalmological research are such motility recordings routinely made. Years ago, cardiologists realized that they could not rely solely on their ears accurately to 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.

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 utility of ERG and VEP, definitive diagnostic motility criteria have been established for all types of nystagmus. The purely clinical signs of these types of nystagmus are often ambiguous and reliance on them without quantitative motility data will continue to cause diagnostic errors that may be compounded by the wrong choices for therapeutic intervention. In many cases, visual acuity can be improved, or binocularity 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 will require the addition of eye movement recordings in the clinical setting to augment the ERG and VEP.

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