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NISTAGMO INFANTILE

I am going to talk about nystagmus of infancy and the presentation will contain six parts. First, I'll define some of these nystagmus types; I'll restrict my discussion to benign types of nystagmus of infancy and discuss some diagnostic criteria that will aid in separating them. Then, I will present some characteristics of each of these types, therapeutic options that are available for some of them, and give some examples.

Definitions and Classifications

The word, «congenital» means, «with birth», but the waveforms recorded from infants where the nystagmus was noted at birth are the same as those seen in infants where they were noted to appear later, after birth. The mechanism for both types are the same, since the waveforms are the same, and congenital nystagmus is equivalent to early onset nystagmus. When babies are born, the whole nervous system is still in development and the moment of birth is no different than the moment before or the moment after. It is a mistake to think of it as a significant event in terms of nervous system development; it is, of course, significant in other ways. Therefore, we need not concern ourselves with whether the nystagmus is truly congenital or acquired in the first two weeks of birth (or a month or two after birth). More important to the physician is, is this a benign nystagmus or is it a nystagmus that is indicative of trouble? Those that have been noted as malignant 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. I am not going to discuss any of those, but we have to distinguish them from the benign type I will discuss.

Congenital nystagmus (CN), latent/manifest latent nystagmus (LMLN), combinations of the two and spasmus nutans — you have to be able to tell them apart. In this classification of CN, sometimes there is a latent component to the CN and sometimes there is one type of the nystagmus blockage syndrome

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(NBS); the other type of NBS is a mixture of CN and LMLN and I will discuss this also. Another time when it is important to distinguish between CN or a benign or malignant nystagmus is when an adult is in post trauma or some other condition and for some reason you cannot get a correct history. Under these conditions, it may be unclear whether the nystagmus was there before or not, especially following trauma. For example, if someone fell in a store, was noted to have nystagmus and then decided to sue someone, you have to be able to determine whether that nystagmus was there before the trauma.

One thing that I should also mention is the associated sensory defects that accompany many types of nystagmus of infancy. We've determined that the etiology and the mechanism of these nystagmus types are independent of those sensory defects and therefore, while there is an association between many sensory defects and the presence of nystagmus, there isn't a causal relationship. The defect does not cause the nystagmus and, vice versa, the nystagmus does not cause the defect. What you have to be careful about is that when there is a sensory defect it, rather than the nystagmus, may be the limiting factor for acuity and even if you halt eye motion using forceps you won't get a big improvement in acuity because the afferent system is defective. If you have a child, or any subject, with nystagmus and you can't correct acuity better than 20/60, you should look carefully for afferent defects. Let me next add a bit in an area that I know little of, genetics. It is my feeling, based on what we've learned, that there could possibly be two different genes with variable probabilities of the one causing the slow eye movement instability resulting in 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. That's one way you might account for the fact that many times there is an afferent defect accompanying CN. Another possibility is just one gene with different probabilities for an afferent defect and CN; you can have both, one or the other. That's all I will say about genetics, but that is the way I envision the causality rather than a direct causal link between these two (CN and an afferent defect); one gene that may cause both or two genes with an association between them.

Benign Types

The benign types of nystagmus are: CN, LMLN, spasmus nutans, the blockage syndrome and combinations of CN and LMLN; this latter is a very small percentage of patients and very difficult because the waveforms are a combination of the two. CN is defined as follows: it can be either pendular or jerk CN, and the pendular CN results from the instability in the slow subsystem resulting in periodic motion of the eyes away from the target and back to the target, not across the target. The waveform looks like a sinusoid but many times it is distorted for reasons we will discuss in a little while. Occasionally there are small

foveating saccades present on the peaks corresponding to where the foveation occours. The jerk forms of CN are caused by an instability that leads to an acceleration of the eyes away from the intended gaze angle or the target and requires a saccade in the opposite direction to stop that runaway. The saccade might return the eyes back to the target or at least start them on the way back to the target for refoveation. We define the direction of the nystagmus by the direction of that saccade despite the fact that we know it is the slow eye movement that causes the nystagmus.

Manifest latent nystagmus, first defined by Kestenbaum I believe, is present with both eyes open but only one being used for fixation. There's always a tropia and 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; the straight eye. Spasmus nutans is «congenital»; it could appear after birth also (a nystagmus of infancy). It is pendular and usually, but not necessarily, ceases by the age of three. It is also usually monocular or disconjugate and is accompanied by a head oscillation many times; we can now tell spasmus nutans from other forms of childhood nystagmus. The blockage syndrome is a very misunderstood syndrome. First of all, the 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 is a strabismus 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. Unfortunately, for the diagnostician and patient who has both CN and LMLN (it is not usual, but some patients have a combination of CN & 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 learn to diagnose the easy types and then worry about the more difficult types. So, how do we diagnose these types of infantile nystagmus?

Diagnostic Criteria

If you are able to record eye movements, the diagnostic criteria for CN are very clear. Any of the twelve waveforms that have been identified in CN are diagnostic of CN except pure pendular; that could be an acquired nystagmus but usually you will find the pendular nystagmus of CN distorted so the patient can foveate whereas in acquired nystagmus you won't see that. In acquired nystagmus, the slow phases can be linear, of increasing velocity, of decreasing velocity or pendular. Whereas CN slow phases can be pendular, increasing velocity or even look linear, when we record MLN it's a decreasing-velocity expo-

nential. We identified, within the pendular or jerk major types, three different forms of pendular and eight jerk; four unidirectional and four bidirectional. One waveform, dual jerk, is a combination of pendular and jerk. I'm not going to discuss each one further; they are available in the literature and I've done it before many times, even here when I last visited Sardinia. If you take a group of patients with CN, 87% will have more than one waveform; they can have any number of the twelve waveforms. There are small percentages of patients with only one (13%, 5% with pendular etc.), but most will have more than one waveform.

The diagnostic criterion for LMLN is also the waveform; it is a decreasing-velocity exponential. All patients (100%) with latent (LN) or manifest latent nystagmus (MLN) have strabismus. I have included in the definition of strabismus, the latent strabismus (phoria) resulting when you cover an eye. Thus, we have a pure LN where the eyes are straight with both eyes open; when you cover one eye, it will develop an eso — or exophoria followed by LN. The waveform has decreasing-velocity slow phases. Pure latent nystagmus i.e., no nystagmus with both eyes open, is very rare, very rare. If you occlude the left eye and the right eye is fixating, jerk-right LN with decreasing-velocity slow phases results. If you occlude the right eye and the left eye is fixating, jerk left with decreasing-velocity slow phases results. Fixating with the left eye, the velocity traces clearly show the jerk-left saccades. Cover the left eye and the right eye, which is usually esotropic, becomes the fixating eye and jerk-right nystagmus results while the left eye becomes esotropic (or esophoric behind the cover). Latent nystagmus implies strabismus.

The diagnostic criteria for spasmus nutans have only recently been defined, in the last couple of years. We can now tell a mother her child has spasmus nutans immediately, by recording; you don't have to wait three or four years before telling her. The waveform is a dissociated pendular nystagmus and the dissociation can even be uniocular. The key is the variable difference between the oscillations in both eyes, unlike congenital and latent nystagmus that are conjugate. The spasmus nutans waveform can vary anywhere from pure conjugacy to pure disconjugacy; 0-180° phase shift. It varies during the recording; you can watch the phase change. When you see that your no longer have to worry about CN; it's spasmus nutans. While we were doing this study we looked at all the patients we had at the time that were very young and we only found three with disconjugate pendular nystagmus that we originally thought had CN. No patients over five year old had disconjugate waveforms; they, we know had CN. The other three have been reclassified as spasmus nutans. Figure 1 is a record from one child with spasmus nutans; the phase shift is different in each segment; purely uniocular, +1 phase (in phase but different amplitudes) and −1 phase (out of phase with different amplitudes). You have to look carefully and follow each cycle in one eye and compare it with the other; if you do that, you'll see the phase relationship. That is how you identify spasmus nutans.

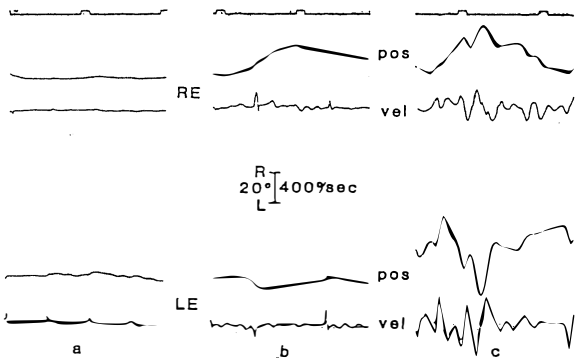

Fig. 1. Example of nystagmus of spasmus nutans with the following relative amplitudes: a) uniocular oscillation of left eye (LE); b) binocular oscillation with no phase difference; and c) binocular oscillation with 180 degree phase difference. RE indicates right eye, R is right, L is left, pos is position, vel is velocity and timing marks represent 1-second intervals in both Figures.

Now, the nystagmus blockage criteria — as I said before, there are two types; in one type CN changes to damped CN, so acuity is better (that's why the patients induce the esotropia). In the other type, the CN becomes LMLN and even though that's a bad waveform for acuity (because there's no long foveation period), it's a very low amplitude and acuity increases. Obviously, to do this, patients must have some sort of variable strabismus. If you are binocular you cannot turn one eye in without getting diplopia; these patients turn one eye in and they don't get diplopia (they suppress that eye); they have a variable, purposive strabismus. When they are looking in the distance they are orthotropic; they become estropic by design and then they either have one or the other low-amplitude waveform along with better acuity. Figure 2 shows a case of the blockage syndrome where there is a CN waveform in the distance (to the left of the Figure is the CN waveform in distance). With esotropia, there emerges a latent nystagmus waveform and the patient sees better because of the long periods where there is little oscillation and this type 2 blockage syndrome switches to LMLN. Kommerell reported a case of type 1 NBS and we've also seen a type 1.

There is a very small group of patients that confuses everybody — CN and LMLN together. Some of them are mostly CN (CN/LMLN) and their waveforms are the CN waveforms and one other waveform called dual jerk latent. They don't have any pure latent waveforms so therefore, CN is predominant. The other group is LMLN/CN because their waveform is latent and dual jerk latent; same dual jerk but the predominant waveform is latent. There are some who have equally CN and LMLN. They sometimes have the CN waveform, other times just the latent waveform and sometimes dual jerk; they exhibit everything. These difficult patients must be recorded to see where they fit; this

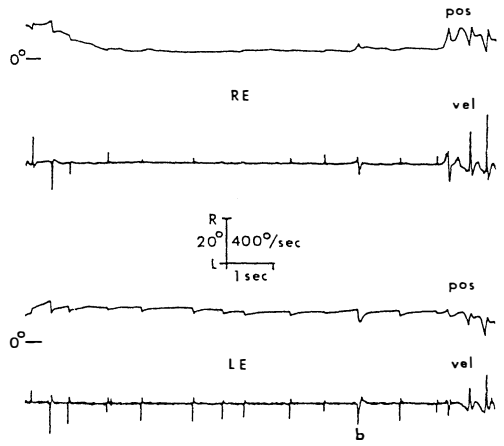

Fig. 2. Example of the nystagmus blockage syndrome. Position (pos) and velocity (vel) recordings of both eyes during binocular viewing of a target 15 degrees to the right (R). The waverforms are CN at the beginning and end of this recording when both eyes were simultaneously on target (i.e., no esotropia). As the right eye (RE) became esotropic, the CN abruptly changed to left-beating MLN which persisted until the esotropia disappeared. The «b» signifies a blink.

group is a very small percentage of patients. A dual jerk waveform is a jerk waveform plus a pendular waveform. A linear slow phase is not diagnostic of anything. But, if the slow phase is accelerating, it is a congenital dual jerk. If the slow phase is decelerating and a pendular is added on, it's a dual jerk latent. You have to look carefully at what is happening on the axis of the pendular slow phase (is it decelerating or accelerating?) and you can then properly categorize the nystagmus. I have developed a classification scheme that I am not going to spend a lot of time on; it was written up in Japan (Japanese Journal of Ophthalmology, 1985) and is available. Basically, you have a large category of pure CN, another large category of pure LMLN and a small category that is a mixture of the two. We have twelve CN waveforms, only one latent waveform and one mixed waveform (dual jerk latent). Thus, we add to our original twelve CN waveforms, just two. In this categorization tree the CN group is I, the LMLN group is II and the combination, III. There are different combinations of the wareforms making subcategories. Patients walk into your office with their eyes wiggling and you have to guess what they have before you record them. How many have CN, how many have LMLN and how many have both? If we look at our first 100 patients without distinguishing between the two, a large percentage (80%) were CN and 15% LMLN wit only a small percentage, mixtures. Thus, if you want to guess, you guess CN. If you look at the

first 100 patients that just had CN, 94% had pure CN (category I) and only 6% had the mixture; they are a very small number of patients. If you look at just LMLN patients, 3/4 of them will just have LMLN, but there's a significant amount of them that will have mixtures. So, more patients with predominantly LMLN will also have some CN than patients with predominantly CN having LMLN.

CHARACTERISTICS

We are now ready to talk about some other characteristics besides the diagnostic waveforms; there are obviously things you can look for in the office. In CN, you should look for a null angle and a clue to that is the child who has a head turn; a teenager or an adult may not because of societal pressures. They' ve learned to keep their head 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.

Convergence null — if patients hold things close to read you can suspect that converging the eyes will null the nustagmus. They may have a latent component; you have to check that by the cover test. If the direction reverses you still don't know whether it's LMLN or CN with a latent component (they are different) but you know that something is happening if it is latent. They may have head nodding, that is not compensatory (I will explain that in a bit). Is there a strabismus or not when the patient has CN? We'll look at that also. CN may show a null angle (the CN magnitude damps if there is a null). The definition of a null (a real null) is that on either side the nystagmus must get larger. I say that because MLN may vary monotonically with gaze angle (Alexander's Law) so that the patient may keep their eyes way to one side where the nystagmus is low; that isn't a true null because there is no way to get an increase on both sides. It's a variation discovered by Alexander when looking at vestibular nystagmus. There has to be an increase on both sides to be a null. Now, the position of this null turns out to be a function of the angle that you are looking at (gaze angle) and it is also a function of the pursuit velocity of the eyes. The null during fixation does not usually equal the null angle during pursuit. As a matter of fact, most times it's shifted in the direction opposite to the pursuit. If you pursue something to the left your null moves to the right of the static null; and if you pursue to the right the null moves to the left. Again, in the first 100 patients (just to get a feel for the numbers) 48% have both convergence and gaze angle nulls, 29% only convergence nulls, 9% only gaze angle nulls and there are 14% with no nulls. We still had to find a way to treat those latter patients. No surgery, no prisms, but Dr. Traccis and I did find a way.

Horizontal head position — the child turns the head to see better. He didn't have to wait to be told about his null, he knows where it is; things get clearer

when he does that. We think of, and I think of, it as a region of ocular motor equilibrium. You have a brain stem (left and right) with forces pulling the eyes both ways and there's a position of equilibrium of forces, not necessarily in primary position, where the nystagmus is minimal. When you find the null, acuity should increase; if there is a severe afferent defect the acuity may not increase.

Tilted head position — sometimes the child will tilt the head; sometimes turn and tilt it. Perhaps the oblique muscles are involved here. This hasn't really been studied well. This represents innervation of muscles other than the horizontal muscles that are somehow helping in reaching equilibrium in what are basically horizontal oscillations. Almost all CN is horizontal, no vertical components. Few patients, there are always a few, have vertical. CN is mostly horizontal and yet, tilting the head is innervating vertical and oblique muscles and that somehow is helping to null a horizontal oscillation. Again, the acuity increases if there are no afferent defects.

Latent components — the CN might reverse direction because when you cover an eye (similar to when the person pursues) the null moves and therefore, you may get a direction reversal of the CN because you go from one side of the null to the other side of the null. MLN mimics latent nystagmus exactly if it is bidirectional. Sometimes it's only unidirectional and when a patient fixates with the left eye perhaps they won't have nystagmus and the right eye will be esotropic but when they fixate with the right eye and the left eye is esotropic, they will have latent nystagmus. That is a curious form; I'm not going to take any time with it. With both eyes uncovered let's assume the null is in primary position to make it easy. To the left, jerk-left CN, to the right, jerk-right CN. If we cover the right eye, the null moves to the right of the primary position so, if you are looking at the person in primary position and you cover the right eye, he would now be to the left of his null which means he would have jerk-left nystagmus. If you cover the left eye, the null shifts to the right so, if you are in primary position, he now has jerk-right nystagmus. It looks just like LN or MLN, but it isn't because the waveform is still CN; it doesn't change waveform it just changes direction because the null has moved with occlusion.

This brings us to the spontaneous head oscillations you see in many children with CN; even adults when are concentraating on something, but adults learn to keep their heads still because it is unacceptable to go around like that. These oscillations use existing pathways in the neck muscles. Quite a number of years ago Dieter Schmidt did EMG in the neck muscles and saw that when normals made saccades to the left the innervation could be seen in the left-turning neck muscles and when they made them to the right they could be seen in the right-turning neck muscles. Normally, in everyday life, when we look left we are going to turn our head left. Those pathways are there so, if there is an instability causing the eyes to oscillate and you measure in the neck, you'll see the same waveform; the EMG will be there. And when the oscillation gets big enough the head will start oscillating. This is not something willed by the pa-

tient to accomplish anything; this is a manifestation of an existing oscillation on existing pathways to the neck. It used to be thought, and some still think, that head oscillations were compensatory. The head was supposed to be moved equally ad oppositely to the CN to stabilize the eye in space. If that were true, you'd need a VOR gain of zero. You can't have a VOR if you are going to move your head to the left when your eyes are moving to the right. With no VOR you would have to move your head in very complex ways. The CN waveforms are very complicated and you have to move your head exactly opposite to get stability; that is clearly impossible. The head has too much mass to duplicate the waveforms of CN. This compensatory hypothesis, when you understand the VOR as you all do now from yesterday's lectures, doesn't work. For realistic compensation, it can be postulated that you could suppress a normal VOR to near zero and only move your head equal and opposite to the movements of the CN during the foveation periods. Perhaps you could achieve stability only during part of the waveform. That is a possible form of compensation.

In most patients with CN, the VOR is normal; it is not something that is affected by CN. The head oscillations are just an extension of the CN and the foveation periods stay flat in space; they don't move and the acuity is unaffected by the head movements. The head oscillations with a normal VOR are equivalent to those of a normal person moving their head. The acuity doesn't go up and it doesn't go down with the head oscillation. In a few patients, the VOR is suppressed and the head moves opposite to the eyes during the foveation periods; the foveation periods become motionless in space because they were not good foveation periods before the head was moving; here, acuity can increase. I have concluded that this is very rare. When there's really no good foveation period, very little cessation of motion, the head oscillates in a way so that during the foveation period it is compensatory and gaze becomes flat. This rare occurrence can occur in some patients with bad CN, CN with no long foveation periods. If patients have a good foveation period there's no reason to shake their head. Thus, if the foveation periods are flat, the head motion cannot help you, you already have flat gaze (no movement during foveation periods) so shaking your head doesn't help you at all. If they are not flat, perhaps some patients can learn to flatten them by moving their head in a certain way.

What is the relation of strabismus to nystagmus (to the CN or the LMLN)? I've already said that LMLN means you have strabismus. Let's, first of all, look at strabismus. Of all strabismus patients, 50% don't have any nystagmus. So you can rule them out for this presentation. Fifty percent do have nystagmus of some type; the question is, what type? It turns out that 26.5% of that 50% of the total will have CN, only 17% will have LMLN and 6% will be mixed. If you take 100 patients that walk in with nystagmus and strabismus and you have to guess (even though all patients with manifest latent nystagmus must have strabismus), your best guess is CN because there are many more CN than LMLN patients and the mathematics works out to favor CN for patients with strabi-

smus. For 100 patients with CN and LMLN, 43% have strabismus and 57% have no strabismus. So, the patient who walks in with nystagmus probably doesn't have strabismus; more than half don't have strabismus. I'll work this out one step at a time. If you take just CN patients of all types, almost 3/4 don't have strabismus. Thus, most CN patients don't have strabismus. If you look at pure CN patients, it's more skewed towards no strabismus. A great majority of them do not have strabismus. We've recorded every CN waveform (all twelve) in patients with binocular alignment, so it is not necessary to have strabismus for any particular CN waveform. Now, this is where I was jumping to. Of 43 strabismus patients in that first 100, more than half had CN even though most patients with CN don't have strabismus. If you have a patient that walks in with strabismus and his eyes are moving, the best guess is CN, a third have LMLN, the rest have mixtures; that's because there's much more CN in the population than LMLN. Strabismus with nystagmus: 53% — CN, 35% — LMLN and 12% — mixed.

The caracteristics of LN and MLN — first of all there's the Alexander's Law threshold, the presence of head turns and, of course, strabismus. Before we look at Alexander's Law, 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 that have pure LN. Many ophthalmologists will tell you they've seen patients with pure LN. When you record them you find it's MLN. If you look with an ophthalmoscope, you can see it. More common is pure LN in primary position with MLN in lateral gaze; most common is MLN at all gaze angles.

Alexander's Law — the instensity of the nystagmus is greatest with gaze towards the direction of the fast phase. Jerk right nystagmus is greater in right than in left gaze. Jerk left nystagmus is greater in left gaze, lesser in right gaze. These are not true nulls, you cannot show increased nystagmus because the patient is at the end of the excursion of the eye. It 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 increasing amplitude (with increasing-velocity exponentials) as you go away from the null in both directions whereas in LMLN (right eye or left eye fixating, you see an Alexander's Law relationship, jerk right greater in right gaze and jerk left greater in left gaze and decreasing-velocity exponentials. That's the difference between the two when you record them. We are not going to talk about other types of nystagmus. The patients with LN usually place the 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; it might go the other way. Again, it minimize the nystagmus and maximizes the acuity. A patient could place his eye in other than the minimum position of nystagmus because he might have what is called an «angle kappa»

that requires eccentric fixation. Now, these are rare, but they exist. They get better acuity despite the fact that the nystagmus might be a little higher where they place their gaze. It can get confusing with MLN as to what head turns really do. We have never recorded the LN waveform in patients with orthophoria; they all had latent strabismus (when you cover one eye, the other doesn't 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 on or out to have MLN. If they can straighten their eyes, the MLN disappears; in the blockage syndrome, they have CN. Therefore, LN and MLN imply strabismus. Of course, strabismus doesn't imply LMLN (50% have no nystagmus at all), but if someone has LN you know they have strabismus. Even if you don't see it clinically (it may be a micro strabismus that can be recorded; we can tell one half degree of strabismus easily) it's there. Strabismus is a necessary, but not sufficient, condition for LMLN. It's either a latent strabismus, if it's pure LN (they're very rare), or a tropia for MLN. CN can occur with or without strabismus; all MLN patients have strabismus.

Spasmus nutans — here, unlike CN, the head nodding is compensatory. The ocular oscillations are asymmetric between the two eyes and, contrary what is written in many books, it doesn't always disappear. You may have patients of 10 or 12 years old that still have spasmus nutans. Many times it disappears to clinical observation (again, like MLN) but if you record it, it's still a pendular dissociated nystagmus that you will find. Now, the head nodding in spasmus nutans is very curious. The VOR of these people is normal, but by shaking their head willfully, the nystagmus is switched off and the eyes become stable in space because of their good VOR; 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. All of a sudden the head starts shaking and the nystagmus stops; due to a normal VOR, the eyes are moving conjugately equally and oppositely to the head, so gaze is now constant. That's how such patients cancel the convergence nystagmus of spasmus nutans, disconjugate pendular oscillation with a still head, conjugate VOR when the head is moving. When you add the eye motion in head and the head motion you wind up with a straight line (gaze is stable) and acuity increases.

The blockage syndrome — they are esotropic at distance. This is something a person with CN cannot do if he doesn't have the ability to partially suppress the eye he purposely makes esotropic. If CN patients with normal binocularity made an eye esotropic they would have diplopia, oscillating diplopia — very bad. These patients can turn one of their eyes in and suppress the esotropic eye; their nystagmus damps and they adopt a head turn to place their viewing eye in adduction. As I said before, the CN is either reduced in amplitude or converted to a low-amplitude LMLN by this purposive of esotropia; the more they increase their esotropia, the lower the nystagmus and as the fixating eye

moves from adduction to abduction the nystagmus increases and esotropia decreases. They usually put their fixating eye in adduction.

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 idea that CN is caused by poor fixation reflexes. We have found fixation in CN is very accurate (< 13 minutes of arc). In LMLN, the fixation reflexes seem to be ineffective since, as soon as a fast phase is made, the slow phase immediately takes the eyes away with high velocity. These patients seem to have no ability to use their fixation system to keep the eye on the target. You can increase the visual acuity of patients 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. Then you can try prisms or surgery but if patients don't have these characteristics, you are basically wasting your time if you want to increase acuity. You might still wish to straighten their head or do some other things, but they must have these characteristics in order to expect an increase in acuity. If you can minimize their attempts to fixate you will maximize their acuity because in CN, the harder they have to try to see (by moving their heads or whatever) the higher the level of nystagmus. Even in turning their head to find the null, the level of nystagmus of that null is higher than if you do the surgery and place the null straight ahead so they don't have to turn their head. Then, the null is better because there is no effort involved. The acuity is going to be proportional to how much time in each cycle they can foveate the target. The longer they can stay on the target the more detail they can get. The retinal slip during this time has to less than five degrees/sec, which is very generous; you'd like to get it less than three or four degrees/sec or acuity will diminish. The data on normals show that when retinal slip stays below four degrees/sec, acuity is the greatest. Also important is the stability of these foveation periods; there is one every cycle. That means the eyes must be in exactly the same position from cycle to cycle; that's called jitter in engineering terms. Jitter must be minimized otherwise, even if they have a long foveation period, one time it will be on the left of the target and the nex time to the right; they are not going to get the detail they need. Thus, all of these must be present in order to have good acuity. How is this done?

For a start, with surgery the surgeon's favorite place to start, one way is the resect and recess operation. Kestenbaum and Anderson independently came upon this. What they do is effectively rotate both eyes opposite to the nulled angle such that the same innervation that used to put the eyes at the null now puts them in primary position and therefore, the nystagmus is nulled. They do-

n't really rotate the eye, they attach the muscles to different places but the way you can think about it is, if the null is 20 degrees left, you rotate the eye 20 degrees right and sew the muscles on. Then, the innervation to look straight ahead is 20 degrees left; that used to be the innervation to get to the null, and still is. Now the null is straight ahead; this is the way I describe it even though that's is not the way the surgery is done. We did some measurements on patients that were operated on by John Flynn, an ophthalmologist who's a scientist also and who wasn't afraid to have the eyes measured both before and *after* surgery. We found that we could plot the shift in gaze that we measured versus the millimeters of surgery that he performed; this was done blind (when I made the measurements I didn't know the extent of the surgery). Later, we took the surgical data from the surgery ward (the operation records) and the points fell along a curve; we dotted in low values. We don't know what these lower values look like, but we presume they are similar to the higher values and we can actually cause the null to shift in towards primary position. The millimeters are the sum of the resection and the recession. If you resected a right lateral five millimeters and recessed the medial rectus five millimeters, the surgical rotation would be ten millimeters. Here's an example — If the null is 20 degrees left, we want to rotate the eyes 20 degrees right. We see from our curve that we need 10 millimeters of total rotation, 5 millimeters per muscle. That is just the way we did it; there are ophthalmologists that use other formulas that I don't think change much; I think half and half is just as good as any other formula and here's how you might do it. Resect the left medial rectus 5 millimeters and the right lateral rectus 5 millimeters, recess the left lateral rectus 5 and the right medial rectus 5 and that will give you a 20 degree rotation.

What if the patient also has strabismus? Not only does he have a null angle, but he has strabismus and you want to perform surgery that will minimize the nystagmus and straighten the eyes. How do you approach that? I think the best way to do this is to operate on the good eye first, the fixating eye, not the strabismic eye (don't worry about it). Correct any remaining tropia after the operation. That means you wait a couple of days and maybe you won't have to do anything because sometimes the surgery you do on the fixating eye reduces or eliminates the tropia. Sometimes you make it worse but it's best to do it that way so that you find out what effect the surgery will have on the tropic eye and then you just move the tropic eye so it's straight; that's not going to affect the nystagmus. So, if the strabismic eye was esotropic and the null was in adduction of the fixating eye, the surgical rotation has to be abducion. That will result in less esotropia. It turns out that this is the most common case. First of all, because esotropia is more common than exotropia and, it turn out, that the combination of esotropia and the null in adduction is more common that esotropia with the null in abduction. This causes in a good surgical result on the tropic eye without touching it. In the other cases you might get more ET or more or less XT. Whatever you get, you then operate on the tropic eye later. An exam-

ple — Suppose preoperatively the left eye was 10 degrees ET when the right eye fixating at 0 degrees and the null was 10 degrees in adduction; you would rotate the right eye 10 degrees. If you rotate the right eye 10 degrees to the right (abduction) then, to look straight ahead, the patient has to look to the left and, in looking to the left, the esotropic eye is going to straighten somewhat (maybe totally) in which case you wind up with straight eyes and a null in primary position. Of course, that's ideal, but that's how it works, not always as perfectly.

How do you access the result of your surgery? This is where some surgeons make a big mistake. Never use head turn — NEVER, NEVER. You have child who realizes his parents are spending a lot of money and who the doctor tells the surgery is going to make his head straight; the last thing you want to do is trust his postoperative head turn on the first visit after the surgery because that child is going to keep his head straight no matter what and thus begins the myth that the surgery was correct (no head turn) and six months later, by magic, the head turn came back. You can't use the head turn because it is under control of the child who is subject to all kinds of input; he wants to please his parents, he wants to please the surgeon. You must measure the null with the head fixed both before the operation and after the operation, so that there is no input from the patient. You have him look at different gaze angles before the surgery and find a null; after the surgery you have to do the same thing; see where the null shifted. When you do it that way, whatever null you measure on day one after the operation you can measure five years later and it will not have moved because it was fixed by the surgery. It is a very important thing to remember that the head turn is useless for evaluating the surgery. In one case, with a very sharp null at 20 degrees left, postoperatively (ten days, four months, one year, we even measured it three and a half years later) the null was still at zero degrees; once surgically shifted, it didn't move. Not only that, but we learned from this that the surgery itself, luckily for the surgeons, broadens the null (so even if they miscalculate the results won't be off by much) and lateral to the null the nystagmus is much less than before surgery. If it was ethical, I would guess that you could cut the muscles and sew them back to the same place and the nystagmus would damp. Just the effect of the surgery reduces the nystagmus, perhaps by scarring where innervation affects the eye; it appears to be a self prophecy that when you operate on CN you get a better result than when you don't, and the null is broader because everything has damped. Therefore, surgery moves the null angle, increases the null area, decreases off-null nystagmus and the new null is stationary. If you didn't do it right the first time it's going to stay wherever you put it; the head turn might vary but the null won't. It may require a second operation. If you miss, if you had a 20 degree null and you brought it in but not enough and there is a new null that's four degrees left, you don't have to do another surgery. You could put prisms in the glasses and move the world to where the null is and fine-tune the surgery with prism. This

is not done very often, possibly because surgery is more remunerative than prisms.

The second way is a bimedial recession. This is artificial divergence; you create an artificial divergence by weakening both medial recti. I would't try this if the patient has strabismus because you are depending upon 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. You weaken both medial recti; you create an artificial divergence and the convergence innervation required to turn the eye to primary position nulls the nystagmus. This, of course, is in a person who has a convergence null; that is the condition under which you would do this operation.

The Faden operation times four means all muscles, all four muscles (by Cüppers). Double vision in lateral gaze is a problem with this operation but some say it doesn't matter because the patient is happy. What they do is weaken all the muscles. If you weaken all the muscles, obviously the result of all innervation is less. 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 is accomplished by tacking the muscle somewhere (5 millimeters) down from its insertion so that you get less force.

Optics - if there is a null to left you can use base-right prisms to move the eyes to the left, into the null. If you measure accurately where the gaze angle null is and the convergence null is, you can translate this directly to prisms or into surgery. If, as I said, you want to move the eyes left, use base-right prisms; if you want to converge the eyes, use base-out prisms; if you want to move left and converge, use compound prisms. Prisms are not (since they decrease fixation attempt) substitutes for head turns: they yield higher acuity than head turns because there is less overall innervation. Feedback of the eye motion from the inside of the iris also may damp CN. We used this effect by inserting soft contact lenses in a subject with CN and decreased the nystagmus in half. Contact lenses with anesthetic (to block afferent input) had no effect on the CN. Thus, soft contact lenses can decrease CN also. How can you tell? Take a cotton swab and just touch the eyelid to see if the nystagmus damps. We are not pressing on the eye we are just touching it a little bit; it's a quick way to tell whether the soft lenses might work. We found no waveform changes and the foveation time increased, therefore, in proportion to the decrease in CN; we think that contact lenses will be beneficial in patients without a null. Even if you don't see damping, it is very easy to try contact lenses. Unfortunately, for many years ophthalmologists have not even prescribed contact lenses for people with nystagmus. They were afraid to put lenses in eyes that were wiggling. It turns out that soft contact lenses at least are not contraindicated by CN and may be indicated as a good therapy. I'm not going to talk about biofeedback,

but it is another option; Ken Ciufredda and Satoshi Ishikawa have written papers on this. I'm not going to talk about acupuncture either; Satoshi Ishikawa has written papers on this also and it's a means for reducing CN.

Examples

That brings us finally to some examples of the different kinds of patients and what you can do. Patients with CN and a null angle allow you to detect its direction, the amount of damping, the foveation time, and you can then choose R&R, soft contact lenses or prisms. You have a choice of therapy for someone who has these nulls. For a person who has convergence nulls (producing damping and increased foveation time) and doesn't have strabismus, you can try base-out vergence prisms with - 1.00 correction, bimedial recession or soft contact lenses. This will work for both nulls, perhaps. If a patient with CN has both nulls, and most with CN do have both, you compare the damping and the foveation time between the null angle and the convergence null. Almost always, the convergence null is the best, better than the version null. Convergence damps the CN more than gaze angle. I would try base-out prisms with - 1.00 or bimedial recession or soft contact lenses. Unusually, when the gaze angle null is better, you do an R&R Kestenbaum or you can use prisms and soft lenses. Suppose there is CN with neither null angle nor convergence null; again soft contact lenses or the Faden operation are possibilities. It doesn't matter if you have a null; if you weaken all the muscles the nystagmus damps.

LMLN - look and see if they follow Alexander's Law; type 1, 2 or 3 should determine where the nystagmus starts damping. If they have Alexander's Law threshold and monocular fixation (they always fix with one eye and the other is always tropic), then they will likely put it in adduction and have a head turn. You can operate on that eye, even though he has strabismus; operate on the fixating eye and move it so that the low-amplitude nystagmus is straight ahead. Then you can correct any remaining tropia just as in CN because the original surgery might change the tropia. Thus, even in latent nystagmus you can perform some operations under suitable conditions. If the patient is an alternate fixer, first he looks with the right eye and then the left eye, and we saw that when he looks with the left eye the right eye is tropic, he'll probably have two head turns. There you don't want to operate, except perhaps the Faden. Certainly no rotation because he alternates his fixation.

Spasmus nutans - it's either going to go away or it's going to decrease to a subclinical level. The therapy is the diagnosis; we can now tell the mother what it is and she can rest easy.

The blockage syndrome - again no therapy other than conventional CN therapy, except possibly bimedial recession. I believe some clinicians are doing bi-

medial recessions on these patients. I don't know the results; it's too early to tell. If it doesn't get published, don't do it.

Finally, combinations of CN and LMLN - the therapy is consistent with all the above constraints; if it's mostly CN you look under CN and what your choices are; if it's mostly LMLN you look under LMLN and see what your choices are.

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