

# READING EYE MOVEMENTS

in

## PATIENTS WITH NYSTAGMUS

and

# PROPOSED THERAPEUTIC PARADIGMS

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### Abstract

*Nystagmus is an involuntary, repetitive, to-and-fro movement of the eyes. It can be divided into two broad categories: jerk and pendular. Patients with nystagmus have decreased visual acuity due to the high velocity retinal-image motion which smears the image of the stationary target being viewed; and, in cases of the congenital idiopathic variety, in addition to any anatomical foveal malformations, they receive bilateral early abnormal visual experience due to this increased retinal-image motion. Reading becomes a difficult task secondary to the error-producing nystagmoid oculomotor component superimposed on the basic reading movements in addition to the reduced visual acuity. We provide a brief overview of the different types of nystagmus, the potential problems that may occur during reading in patients with nystagmus, and treatment options that may be helpful in such cases.*

### Key Words

reading, jerk nystagmus, pendular nystagmus, nystagmus vision therapy, eye movements

**R**eading is a complex, higher-level process which requires the oculomotor system to perform optimally and efficiently, frequently for extended periods of time. Reading becomes difficult in patients with nystagmus because of the error-producing abnormal nystagmoid oculomotor component added to the basic reading process over and above the reduced visual acuity. It is important for the clinician to understand the basic characteristics of nystagmus and how it may affect reading efficiency, as well as the range of treatment modalities available to such patients.

### Nystagmus Overview

Nystagmus is an involuntary, repetitive, to-and-fro movement of the eyes. Classically, nystagmus is divided into two broad categories: pendular and jerk.<sup>1,2</sup> Either can present in patients having a congenital or acquired etiology.

Pendular nystagmus is characteristically described by smooth horizontal or vertical sinusoidal movements, generally bilateral and relatively equal in velocity in both directions. The dynamic characteristics of pendular nystagmus may vary. Typically it has an amplitude of one to eight degrees, a frequency of two to six cycles per second, and a peak velocity of up to 100 degrees per second. Pendular nystagmus can sometimes have a jerk component with the fast phase occurring in the direction of extreme gaze.

Jerk nystagmus can be horizontal, vertical, or cyclorotary and is generally bilateral. It is characterized by a slow phase in one direction and a fast, saccadic phase in the opposite direction. The slow phase represents the pathological error-produc-

ing component, while the fast phase represents the corrective foveating response. The amplitude of jerk nystagmus is typically one to five degrees, with a frequency of one to five cycles per second and a slow-phase peak velocity of up to 80 degrees per second. Jerk nystagmus is defined by the direction of the fast phase. For example, jerk nystagmus with the fast saccadic phase toward the left is termed left jerk nystagmus.

There are three basic types of jerk nystagmus exhibiting distinct dynamic characteristics.<sup>1</sup> (See Figure 1.)

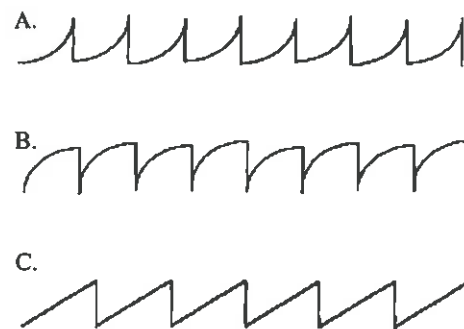


Figure 1. Types of jerk nystagmus. A. Increasing exponential. B. Decreasing exponential. C. Linear. Slight jaggedness of the simulated waveforms is due to resolution limitation of the computer-scanned images.

The first type of jerk nystagmus has an exponentially-increasing, slow-phase component. Simple congenital, idiopathic nystagmus is frequently of this form. The second type has an exponentially-decreasing, slow-phase component. Manifest-late nystagmus is an example of this second type and is associated with strabismus. The third type of jerk nystagmus has

a linear, slow-phase component. It can result from either a central or peripheral vestibular nervous system dysfunction. It is also found during irrigation of the external auditory canals.

Both pendular and jerk nystagmus have a position of gaze where the amplitude and frequency is at a minimum, termed the null point. Thus, when nystagmats attempt to see more clearly an object that lies in front of them, they will frequently turn their head in the direction opposite to that of their null position. In this manner the extraocular motor innervation is similar to that found with the head straight and the eyes directed laterally into the null position.

The velocity of the nystagmus contributes to the reduction in visual acuity found in these patients because of the increased retinal-image motion producing a smeared image. The constantly varying eccentric retinal locus may also contribute to the loss, but this factor has rarely been considered. The nystagmus presents a problem during reading, especially with smaller-sized print.<sup>3</sup> It drives the eyes away from the word of interest, thus interfering with oculomotor positional and text processing. In cases of congenital idiopathic nystagmus, these sensory and motor-based aspects are superimposed on the bilateral amblyogenic sensory component due to early abnormal visual experience as a result of the increased retinal-image motion. Additionally, there are often anatomical foveal malformations. And, in cases of acquired etiology, the nystagmus produces oscillopsia, an illusory sense of movement of the world that is extremely disturbing to a patient. Thus, the effect nystagmus has on a patient will vary considerably depending on the etiology.

### Normal Reading

When reading, one maintains a complex level of oculomotor, sensory, cognitive, and linguistic activity. The oculomotor component involves the interaction of three primary types of eye movements.<sup>3</sup>

The first eye movement is the saccade, which is the fastest of the eye movements. Its peak velocity is proportional to the size of the eye movement; the larger the movement, the higher the peak velocity.<sup>1</sup> There are three categories of saccades that are present during reading. The first type is the progressive saccade, which moves the eyes from left to right over the words in a line of text. The second is the regressive saccade, which moves the eyes from right

to left to reread a portion of text. And the last type is the return-sweep saccade, which is executed near the end of the line of text and moves the eyes from right to left to near the beginning of the next line.

The second eye movement is disparity (fusional) vergence, which typically has a small amplitude of about 0.3 degrees. Disparity vergence is a reflexive, binocularly-corrective response following a saccade during the 200-300 ms fixation pause. It is compensatory for the transient dynamic divergence which occurs during each brief (e.g., 29-45 msec) saccade.

The third eye movement refers to the fixational component itself. When fixating an object, the microsaccadic eye positional and drift velocity errors must be kept at a minimum to maintain maximum foveal visual acuity. This is assured in normals by the relatively brief fixation durations and the heightened attentional aspect. Absence of any relatively large oculomotor errors allows for more rapid and efficient text and positional processing.

Other oculomotor systems may also be activated during reading.<sup>1,3</sup> The vestibular system with its very short latency (about 15 msec) compensates for head movements to maintain steady fixation on a word with maintained clarity of vision. The pursuit system may be activated to compensate for any smooth movement of the reading material itself; for example, slight slippage may occur while reading on a bus or subway. It may also be activated to compensate for the small fixational drift velocity errors, as mentioned earlier. And the optokinetic system may be stimulated by the surrounding Gibsonian visual flow patterns when reading in a moving vehicle. However, to maintain steady fixation, the optokinetic response must be suppressed.

The reading rate of an average college student ranges from 200 to 350 words per minute with good comprehension.<sup>4,5</sup> Reading rate is related to level of difficulty of the text material. Reading difficult material results in a slower reading rate, while reading easier text can result in reading rates up to 500 or so words per minute.

### Reading and Nystagmus

When objectively measuring eye movements during normal reading, one observes a staircase pattern of alternating saccades and fixation pauses.<sup>1,3,4,6-8</sup> (See Figure 2.)

It represents the integration of all progressive and regressive saccades, return-

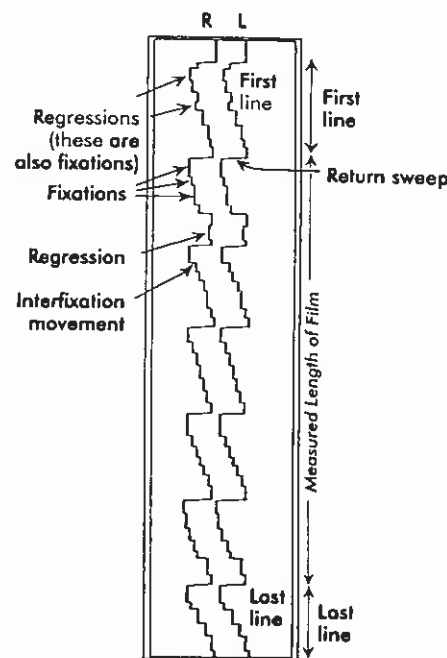


Figure 2. Normal schematic binocular reading eye movement record.<sup>5</sup>

sweep saccades, and fixation pauses (as well as the small and transient vergence movements during and immediately following each saccade). Under typical test conditions, the other possible oculomotor components play a relatively minor role.

Nystagmats also display a similar staircase pattern. However, the nystagmus is now superimposed upon the reading record.<sup>3,4,6-8</sup> (See Figure 3.)

One can see how the presence of nystagmus will cause a problem when reading (as described earlier), which in itself is a complex process. For example, nystagmats appear to make smaller and more frequent progressive saccades, tend to lose their place, and may have difficulty shifting to the next line of print. Hence, reading rate in nystagmats is typically reduced by 20-60 percent when compared with normals.<sup>3,4,6-8</sup> In some unusual cases,<sup>7</sup> however, the nystagmus is considerably reduced during reading. Perhaps the requisite reading-related saccades are able to suppress the jerk nystagmus-related saccades since only four or five saccades can typically be executed per second.<sup>1</sup> The clinician must be aware of this when considering possible therapeutic interventions, as well as discussing potential problems and limitations regarding vocational and avocational aspects, especially with the younger patient.

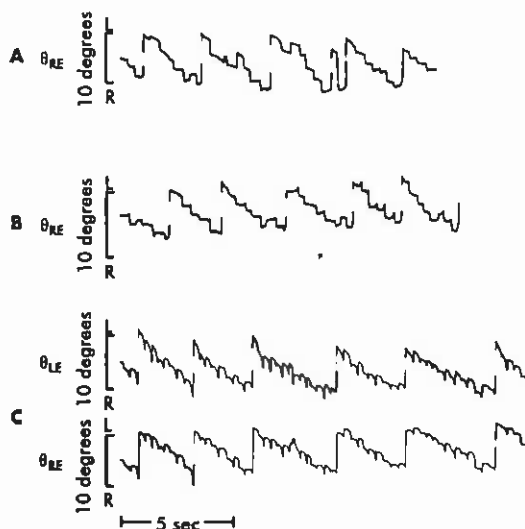


Figure 3. Reading eye movements showing variability of jerk nystagmus and its influence on reading ability. Reading material level (high school) was similar for each trial. Top trace: relatively normal record except for the presence of some nystagmus slow phases (to the right); reading rate 130 wpm. Middle trace: relatively normal record except for the presence of some nystagmus slow phases (to the left); reading rate 156 wpm. Bottom traces: marked nystagmus now present and superimposed on the binocular reading record; reading rate reduced to 115 wpm.<sup>7</sup>

### Proposed Therapeutic Paradigms

There are two broad categories of treatment options available to reduce the amount of nystagmus: traditional and non-traditional. Traditional treatment of nystagmus includes vision therapy, base-out prisms, contact lenses, eye muscle surgery, and pharmacological intervention. Nontraditional treatments include oculomotor auditory biofeedback and rapid serial visual processing.

### TRADITIONAL TREATMENT

#### Vision Therapy

Vision therapy can be used to enhance basic oculomotor skills. Reading efficiency can be improved with basic version, vergence, and accommodative training.<sup>4,9-10</sup> Afterimages have also been used to provide direct visual feedback regarding accuracy of oculomotor fixational and tracking ability, as the foveally-placed afterimage is stabilized retino-cortically,<sup>11</sup> so that the patient visualizes and "sees" the nystagmus. In addition, controlled reading, using variable-speed apertures superimposed over text materials, has been used to improve reading ability by deconditioning regressive movements.<sup>4</sup> Tachistoscopic training has also been employed to increase the span of perception and decrease processing time.<sup>4</sup> In general, the results have been positive and promising,

although a much larger group of nystagmats is needed, probably in the context of a clinical trial, to determine in a more generalized manner the therapeutic outcome following such varied forms of vision therapy.

#### Base-Out Prisms

Use of base-out prisms is probably the simplest approach in the management of nystagmus in the absence of strabismus.<sup>12-14</sup> Spectacles with equally divided amounts of base-out prisms before each eye stimulate convergence, which dampens the magnitude of the nystagmus (probably by interaction of the nystagmus and convergence oculomotor signals). It thereby reduces the retinal-image motion and increases text visibility. This is one reason why young nystagmus patients with ample accommodation tend to hold reading material at a closer working distance than normal; they stimulate convergence and reap the visual benefits. Composite prisms (i.e., base-out prisms of unequal amounts before each eye) have been used to

create the same oculomotor innervational pattern in the "straight ahead" position as that found in the null position, with the added benefit of the simultaneous convergence innervation.<sup>14</sup> Some clinicians believe that vision therapy should first be implemented, and only then prisms considered as a supplement, in all patients who show improvement during testing with prisms.<sup>15</sup> However this philosophy remains a point of debate.

#### Contact Lenses

Rigid gas permeable lenses may also help patients with nystagmus. Contact lenses provide a wider field of view which may be beneficial to fusion and, in cases of high refractive error (especially the higher degrees of astigmatism typically found in these patients), can improve visual acuity by providing better optics than a spectacle prescription. Contact lenses may also reduce aniseikonia in cases of refractive anisometropia, and thus once again promote fusional ability in an already compromised system with its multiple sensory and motor disturbances. In addition, there is tactile feedback when nystagmats wear rigid gas permeable lenses.<sup>16,17</sup> As nystagmus intensity increases, there is greater sensation of lens awareness with changing eye position as the rigid lens moves more vigorously

against the sensitive palpebral conjunctiva. Patients fitted with rigid lenses are able to use the correlated lid tactile feedback to control and reduce their nystagmus in the normal environment once they are made aware of this novel source of oculomotor feedback. We feel that rigid contact lenses are a good supplemental treatment to be used in conjunction with vision therapy and oculomotor auditory biofeedback.

#### Eye Muscle Surgery

Eye muscle surgery is another possible mode of treatment in the management of nystagmus,<sup>18-21</sup> although diplopia may result, especially in the adult patient. Improvement of visual acuity has been found following extraocular muscle surgery consisting of the Anderson-Kestenbaum procedure as well as the artificial divergence procedure.<sup>20,21</sup> The former procedure mechanically shifts the position of the eccentric null point to the "straight ahead" position, thus reducing the need for a head turn to improve vision. In contrast, the latter procedure creates a mechanical situation requiring convergence innervation at distance. Since convergence at near dampens the magnitude of nystagmus, then, surgically causing the binocular motor system to exert convergence innervation at distance results in a similar dampening effect. Post-operatively, patients demonstrated up to a two-line increase in Snellen visual acuity.<sup>21</sup> In addition, if this is done at any early age (before 6 to 8 years), it may reduce the deleterious amblyogenic effects of the increased retinal-image motion. Subsequent active optometric vision therapy in such patients, by treating them as if they were bilateral amblyopes, should yield yet further improvement of overall vision function. Treatment of nystagmats in this way has been given little formal attention, and it would require considerable cooperation between the optometrist, ophthalmologist, and patient/parent.

#### Pharmacological Intervention

More recently, pharmacological agents injected directly into the extraocular muscles have been used to treat patients with nystagmus. Botulinum toxin A is used to paralyze to varying extents the extraocular muscles, thereby effectively reducing the magnitude of nystagmus.<sup>22</sup> It affects the neuromuscular junction by chemodenervation by preventing the release of acetylcholine. However, various side effects, such as diplopia and ptosis as well as forced innervational motor adap-

tive changes, may limit its use.<sup>22</sup> Baclofen has been successful in the treatment of some forms of acquired nystagmus. Its specific mechanism of action is not fully understood; however, it inhibits monosynaptic and polysynaptic transmission at the spinal cord and also depresses the central nervous system.<sup>23</sup> However, these effects may be short-lived, and thus a more permanent pharmacological agent in the successful treatment of nystagmus remains to be found.

### **NON-TRADITIONAL TREATMENT Auditory and Associated Biofeedback Techniques**

The use of oculomotor auditory biofeedback was first reported nearly two decades ago in the treatment of patients with congenital nystagmus.<sup>24-29</sup> The infrared limbal reflection technique was used to monitor changes in horizontal eye position.<sup>1</sup> The reflected light detected by the sensors varies as a function of eye position. These changes are converted to an auditory signal whose pitch alters systematically with eye position. Thus, patients actually "hear" their nystagmus. While the patient fixates a target in primary position, the nystagmoid eye movement is represented as a jerky or wavering tone. The patient attempts to reduce this varying and uneven tonal quality by developing and invoking one or more higher level control strategies. Different strategies may be used, such as visual imagery and relaxation, to gain control over the nystagmus. For example, while fixating a target at near in primary position, the patient is initially instructed to relax and imagine looking "through" the target and display screen into the distance. As the patient relaxes and begins to exert some control, the tone becomes more constant in pitch as the nystagmus is reduced. The patient may then use additional internal control strategies to reduce the tonal variation to a minimum. For example, some patients imagine that they are gazing into their null position, while others imagine looking with either one eye or the other during binocular viewing of the target. The ultimate goal is to reduce the tone to a constant, unwavering pitch, as extending foveation time will optimize visual acuity. The patient is then gradually "weaned" off the tone until eventually control can be consistently exerted at will without auditory assistance under more naturalistic conditions and in the presence of purpose-

ful distractions. These techniques can then be applied to the patient's own external environment, such as the classroom, to increase visual acuity at distance on the blackboard and to enhance text visibility at near as needed for short periods of time (up to two minutes or so), thus effectively functioning as a "spotting" device. At home, the patient is asked to practice these techniques and to monitor the oculomotor variations either directly by tactile feedback (lightly touching the eyelid with a finger) or by observing changes in very low contrast test targets. Also, family members may provide verbal feedback regarding the nystagmus intensity during the home training periods.

Over the past 20 years, we have had remarkable success with this technique. With ongoing developments in this area, including relatively simple and inexpensive oculomotor auditory biofeedback devices for in-office and home training, this potent conditioning paradigm should become more commonplace.

### **Rapid Serial Visual Processing (RSVP)**

The primary way to read more rapidly and efficiently is to minimize the number of saccadic eye movements. The maximum number of saccades made is controlled by the neurological sampled-data system, which limits it to four or five per second.<sup>1</sup> Thus, during simple positional tracking, its 180 ms or so latency/refractory (fixational) period is activated; however, during reading, an additional 70 ms or so is used for text processing during each 250 ms fixation pause.<sup>1,3</sup> Elimination of saccades during reading can therefore effectively result in a savings (~75%) of at least 180 ms per saccade, in addition to the 20 to 45 msec saccade duration itself. However, even with the elimination of the saccades related to reading, the nystagmus component is still present in these patients. Therefore, it is first necessary to use other modes of treatment to improve oculomotor control and enhance text visibility, and then use additional techniques, such as the RSVP methodology, to increase reading speed even further.

Basically, rapid serial visual processing, or RSVP, refers to a computer-controlled text with a fixed spatial location within which text is presented one word at a time.<sup>30</sup> Since saccades are not required during such RSVP reading, it can be a useful tool for those patients who have already attained good fixational control

over their nystagmus, therefore increasing reading efficiency and reading rate even more. RSVP can also be used in conjunction with oculomotor auditory biofeedback to maintain improved gaze stability for longer periods of time.

### **Discussion**

It is clear that nystagmus control is essential for more rapid and efficient reading in these patients. The act of reading requires a sustained and high level of cognitive, linguistic, sensory, and oculomotor processing and interaction. It is easy to see how the abnormal nystagmoid oculomotor component can cause havoc to the complex reading process. The staircase pattern found in normal reading is also present in patients with nystagmus, but with the aberrant nystagmoid eye movements superimposed, overall reading efficiency is reduced.

Our present training protocol in a patient with nystagmus has multiple sequential components which may take several months to complete. First, we conduct eight sessions of eye movement auditory biofeedback. Most of this involves midline fixation; the latter sessions include some saccadic and pursuit tracking. Once relatively consistent control has been established with these initial sessions, additional sources of feedback are incorporated concurrent with the ongoing clinical auditory biofeedback sessions. This includes visual feedback using computer tracking tasks and the pegboard rotator. It also includes tactile feedback involving placement of the patient's index finger against the upper lid. During these times, they attempt to invoke the higher-level control strategies learned previously with the auditory biofeedback training. We plan to incorporate afterimage training in the future, using a device that helps sustain the percept for up to five minutes. At this time, some of the patients are also fit with RGP contact lenses, which typically improves their vision as well as provides an additional source of tactile feedback related to their nystagmoid movements. Then home training is instituted. The patient can use tactile feedback from a finger and/or contact lenses, verbal feedback from a family member trained to detect and assess the patient's eye movements, and visual feedback using very low contrast stimuli (near threshold) such as sinusoidal grating patterns. In this last case, when the nystagmus worsens, the grating pattern disappears, whereas if the

nystagmus decreases, the pattern becomes visible. In the future, we anticipate having a simple eye movement auditory biofeedback device for home training, which will provide reinforcement of the clinical sessions.

It is important for the clinician to consider all viable options in the treatment and overall management of the patient with nystagmus. Whether such treatment is invasive or non-invasive, traditional or non-traditional in nature, or some combination, it is the responsibility of the clinician to educate the patient regarding all interventions as well as any potential problems or side effects. With the variety of treatment options available, as well as others currently being investigated, including more experimental modes such as acupuncture<sup>31</sup> and afferent stimulation,<sup>32,33</sup> the clinician and patient should be able to formulate an appropriate and beneficial rehabilitative program.

*(Supported in part by a grant from the Schnurmacher Institute for Vision Sciences at SUNY/State College of Optometry, New York City.)*

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## EDITORIAL continued

Further, the present system of health care reimbursement is not conducive to healing the patient. Managed care seeks to reduce the number of procedures performed and minimize the time the doctor spends with the patient. Consequently, the doctor's ability to fully understand the disease in order to fully treat, and to gain the knowledge and insights about the patient as a person that are prerequisites for healing, are both compromised. This is the antithesis of Galland's method of "patient centered diagnosis" and "integrated medicine."

Finally, a system of integrated optometry would require the doctor to at once balance a superb knowledge of the diseases that afflict his/her patients with the knowledge and sensitivity to understand them as unique individuals. This means that the *healer* must be doctor, psychologist and clergyperson all at once, indeed a challenging task. Perhaps Galland's system is utopian, yet it is reminiscent of the old time family doctor. Hopefully, what goes around will come around in the near future.

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Date accepted for publication:  
June 24, 1997