# The Benefits of Extraocular Muscle Surgery in Infantile Nystagmus Syndrome

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**OMLAB Report #123007**
**Written:** 12/28/07; **Placed on Web Page:** 12/30/07; **Last Modified:** 8/28/09
**Downloaded from:** OMLAB.ORG
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This work was supported in part by the Office of Research and Development, Medical Research Service, Department of Veterans Affairs.

The following summary is written in response to the reports of patients whose physicians have made uninformed or erroneous comments to them regarding the efficacy of extraocular muscle (EOM) surgery for the improvement of visual function in patients with infantile nystagmus syndrome (INS). Also included are the results of studies demonstrating that the four-muscle tenotomy and reattachment (T&R) procedure is therapeutically effective for both pendular and jerk types of acquired nystagmus. Because the primary purpose of this report is to better inform INS patients (and their parents) and, when necessary, to help them educate their physicians, it is written with both layman and professional in mind.

Patients have reported on the American Nystagmus Network (ANN) and elsewhere that their doctor told them that EOM surgery for INS: 1) does not help the patient to see better; 2) does not damp (reduce the amplitude of) INS; 3) should not be done unless there is a large (i.e., 30° or greater) head turn; 4) should not be done early (e.g., before the age of 4 or 5); or 5) is "controversial." If your doctor makes any of the above (or similar) statements, show him/her the relevant section below. If they are not open to learning the facts, seek a more informed opinion. With regard to the claim that either of the EOM surgeries discussed below is somehow "controversial," there is no scientifically valid foundation for such a claim. Any controversy that does exist is purely political and reflects either ignorance of the literature or a failure to understand the powerful influences that proprioception exerts over eye movements; in some cases it represents bias. One physician stated that he had never heard of either the T&R procedure or one of its leading proponents and "if it was tested and verified it would have appeared in the literature"—where has this "professional" been for the last 8 years?

**Bimedial Recession (BMR or "Artificial Divergence") Surgery (Two-Muscle Recession)**

1. *Only* for binocular (no strabismus) INS patients with convergence damping

Eye-movement data have shown that for these patients, whether or not they also have a gaze-angle null, the act of convergence has the most beneficial effects on INS waveforms. This nystagmus surgery is performed in the same manner as the bimedial recession surgery to correct esotropia. However, its therapeutic effects on visual function in INS are drastically different (1).

2. Improves INS waveforms (i.e., better foveation periods = better visual acuity)

Despite the many reports over the past 50 years of better acuity post-EOM surgery, some are apparently unaware of this important improvement in visual function. Visual acuity improvements were documented in the first ocular motor study of the Kestenbaum surgery (see below) EOM (2).

3. Broadens the range of gaze angles with better foveation (i.e., better visual acuity)

This underappreciated improvement in visual function is not even measured in the physician's office. Broadening the NAFX peak is, in my opinion as both a scientist and a person with INS, more important than a small improvement in primary-position visual acuity. Having a sharp NAFX peak is like having your vision limited by a translucent plastic bag on your head in which a small hole is placed at the angle of the NAFX peak. That necessitates having to constantly turn the head to see clearly anything of interest in your visual field, a time-consuming and stress-inducing requirement that drastically reduces visual function. Broadening the NAFX peak effectively enlarges the hole, allowing eye-movements alone to find and identify targets more quickly over a broad range of gaze angles (i.e., a large part of your visual field).

4. Damps INS (usually damps INS enough to be visible to the naked eye)

Patients and their parents (as well as their physicians) should realize that damping INS is mainly cosmetic; it does not necessarily improve visual acuity. Also, the post-surgical damping may not be noticeable to either the parents or the physician but eye-movement recordings will document the extent of the damping. Although damping will be appreciated cosmetically if large enough, even a small amount of damping allows the fixation system to prolong and steady the all-important foveation periods in every INS cycle. That will improve visual function.

**Kestenbaum Surgery (Four-Muscle Resection-Recession)**

1. Removes head turn by moving the "null" position to straight ahead

The idea that the Kestenbaum procedure should only be done for large head turns (e.g., 30° or more) is based on consideration of only the orthopaedic problems that a head turn might cause. Such an overly restrictive position ignores the demonstrated ability of the Kestenbaum procedure to lessen the severe deficits in visual function caused by a lateral, narrow range of gaze angles where high acuity is possible (2). Instead of the "null" position, a better measure of the INS waveform and its relationship to visual acuity is given by the peak value of the NAFX, a mathematical function that is directly proportional to the maximum visual acuity possible for each waveform recorded (3). Therefore, the Kestenbaum procedure should be used to improve visual function on all patients with a lateral NAFX peak, regardless of the amount it is lateral to primary

position and regardless of whether it causes a consistent head turn; for very small lateral NAFX-peak angles, the four-muscle T&R procedure (see below) will broaden the peak enough to remove the lateral peak and any head turn it might have caused; for moderate lateral NAFX-peak angles, the Anderson + T&R procedure can be substituted for the Kestenbaum procedure.

2. Improves INS waveforms (i.e., better foveation periods = better visual acuity) (see BMR)

3. Broadens the range of gaze angles with better foveation (i.e., better visual acuity) (see BMR)

This underappreciated improvement in visual function is not even measured in the physician's office. Broadening the NAFX peak is, in my opinion as both a scientist and a person with INS, more important than a small improvement in primary-position visual acuity. Having a sharp NAFX peak is like having your vision limited by a translucent plastic bag on your head in which a small hole is placed at the angle of the NAFX peak. That necessitates having to constantly turn the head to see clearly anything of interest in your visual field, a time-consuming and stress-inducing requirement that drastically reduces visual function. Broadening the NAFX peak effectively enlarges the hole, allowing eye-movements alone to find and identify targets more quickly over a broad range of gaze angles (i.e., a large part of your visual field).

4. Damps INS (may or may not damp INS enough to be visible to the naked eye) (see BMR)

Patients and their parents (as well as their physicians) should realize that damping INS is mainly cosmetic; it does not necessarily improve visual acuity. Also, the post-surgical damping may not be noticeable to either the parents or the physician but eye-movement recordings will document the extent of the damping. Although damping will be appreciated cosmetically if large enough, even a small amount of damping allows the fixation system to prolong and steady the all-important foveation periods in every INS cycle. That will improve visual function.

**Anderson + T&R Surgery (Four-Muscle: Two Recessions + Two T&R)**

1. Removes smaller head turns by moving the "null" position to straight ahead (see Kestenbaum)

This is a four-muscle nystagmus surgery despite recessing only 2 conjugate muscle insertions. The addition of a 2-muscle T&R to the original 2-muscle Anderson recessions allows the advantages of a 4-muscle T&R to be realized. There are currently no eye-movement data to suggest that the 2-muscle Anderson procedure alone can duplicate the therapeutic effectiveness of a 4-muscle procedure.

2. Improves INS waveforms (i.e., better foveation periods = better visual acuity) (see BMR)

Anderson may have been the first to recognize that EOM surgery for INS could improve visual acuity (4); this was later recognized in a study of the Kestenbaum procedure (2).

3. Broadens the range of gaze angles with better foveation (i.e., better visual acuity) ahead (see BMR)

4. Damps INS (may or may not damp INS enough to be visible to the naked eye) (see BMR)

**Tenotomy and Reattachment (T&R) Surgery (Four-Muscle Tenotomy-Resuture)**

1. Improves INS waveforms (i.e., better foveation periods = better visual acuity)

This is the purest form of nystagmus surgery, as it does not move any muscle insertions. Instead, it relies solely on the neurophysiological changes in the responsiveness of the eyes to the nystagmus signal from the brain. It does this by using proprioceptive control to reduce the "gain" of the ocular motor plant (i.e., the eye). That means that the eye will shake less even though the nystagmus signal did not change. When there is a sharp NAFX peak in primary position, or even if there is no peak, the T&R procedure will improve the INS waveform and, with it, visual function.

2. Broadens the range of gaze angles with better foveation (i.e., better visual acuity) (see BMR)

As explained above for the BMR procedure, it is the obligate tenotomies in that procedure that were recognized as being responsible for the "null" broadening in the 1979 eye-movement study of the Kestenbaum procedure (2). That key observation led to the development of the T&R procedure on a canine with INS (5); that was followed by the masked-data clinical trial of T&R; that was the first such trial of any EOM surgery (6,7).

3. Damps INS (may or may not damp INS enough to be visible to the naked eye) (see BMR)

4. Improves visual recognition time of new or otherwise displaced targets

Recent studies showed that INS results in longer than normal target acquisition times (the time to move the eyes and fixate on something new); that greatly diminishes visual function in many every-day activities, both professional and athletic (8). The four-muscle T&R procedure has recently been found to significantly reduce this prolonged target acquisition time, thereby improving visual function in a dynamic manner in addition to the static improvements discussed above (9).

In summary, the Kestenbaum, Anderson + T&R, and T&R procedures are four-muscle procedures that improve visual function in both static and dynamic ways; the BMR, although a two-muscle procedure, accomplishes the same therapeutic effects by affecting the proprioceptive tension-control loop in a similar manner. These effects have been demonstrated in peer-reviewed papers in highly respected scientific and clinical journals; there is no controversy about that data. Just as early strabismus surgery can aid in the development of stereoscopic vision, early EOM surgery can aid in the development of visual function in general.

**Maximal Recession Surgery (Four-Muscle Large Recessions)**

1. *Not* recommended in general and *never* recommended for binocular INS patients.

This surgery, conceived in Italy (10,11) and used for a time in Germany before its introduction to the United States (12,13), is a poorly thought out, overly aggressive attempt to beat nature into submission. It is the antithesis of T&R, which maintains homeostasis.

2. Temporarily reduces INS amplitude and possibly improves INS waveforms (i.e., better foveation periods = better visual acuity)

The temporary improvements are probably due to the mandatory 4-muscle T&R that is part of this procedure. Unfortunately, by reducing the saccadic gain in addition to that of the INS slow phases, the brain's plasticity acts to restore normal saccadic gain and, in doing so, also raises the slow-phase gain so that the INS amplitude also increases. The dangers of lateral-gaze diplopia (first recognized in Germany) make this a procedure whose temporary benefits are exceeded by its undesirable side effects.

3. Presumably, temporarily broadens the range of gaze angles with better foveation (i.e., better visual acuity) ahead (see BMR and "2")

4. Presumably, temporarily damps INS (may or may not damp INS enough to be visible to the naked eye) (see BMR and "2")

**Sources of the So-called "Controversy" over EOM Surgery**

1. Failure to understand how EOM surgeries work (i.e., how proprioception reduces the INS by reducing the responsiveness of the eye to the unchanged INS signal from the brain).

For decades ophthalmologists have been trained to "move" muscles in EOM surgery. This has its roots in strabismus surgery, where one strives to correct a misalignment between the eyes. However, in nystagmus surgery, that is not the goal. Rather, it is to improve the INS waveform and, if not already best in primary position, to move the best waveform to primary position. Improving the waveform is accomplished in nystagmus (not strabismus) surgery by changing the proprioceptively controlled EOM tension to make the eye less responsive to the nystagmus signal from the brain (14). Moving the best waveform to primary position is still accomplished by moving certain muscles to reposition the peak NAFX value to primary position. Current ophthalmological training has not yet incorporated these new concepts into the curriculum of ophthalmology residents and it has been difficult for some physicians to understand and accept them. That difficulty has been expressed as "disbelief" in the proven benefits of EOM surgery that improve visual function in several measurable ways. It has also led some to uncritically accept statements in the overreaching abstracts of two scientifically flawed and ethically challenged papers. These papers were surreptitiously conducted and submitted to be published at the same time as the successful results of the masked-data clinical trial of T&R; therefore, the papers violated both the established protocol of the clinical trial and the explicit agreements made with one of its key personnel (see below). Because of the success of T&R in treating INS and both pendular and jerk forms of acquired nystagmus (15,16), the importance of

proprioception in the control of eye movements is just now beginning to be appreciated by ocular motor scientists. However, physicians have used it for over 50 years in the treatments for INS (often without understanding the mechanism, e.g., contact lenses, afferent stimulation, induced convergence, EOM surgeries) (17).

2. Failure to understand the serious scientific faults in a pair of papers written by two people with little or no experience or knowledge of INS data-recording techniques, data-calibration techniques, research analysis methods, or patients.

The history of how the data from the clinical trial of T&R were distributed to others with neither the knowledge nor approval of all the scientists responsible for the clinical trial is treated in detail elsewhere (18). However, even if those responsible for this attempt to subvert the clinical trial had not violated both the letter and spirit of ethical scientific conduct, they were so inexperienced and ignorant of the necessary procedures to take, calibrate, and analyze nystagmus data from INS patients that they made many mistakes in their papers. These too, have been treated in detail elsewhere (19). The bottom line is, false claims were made in the abstracts that could not be supported by the data in the papers, even if the authors had not made fatal methodological mistakes. As mentioned above, some with less than scientific rigor merely parroted those errors in the abstracts without fully understanding either the unproven methods used in the papers or the errors in the application of those methods. That is not the basis for a scientific "controversy;" it is merely fodder for politically driven bias. Every published study of the effects of T&R conducted by experts or other credible scientists in this field of study has documented its positive therapeutic effects that improve visual function; no negative effects of this muscle sparing procedure have been found. If anyone uses these tainted and scientifically flawed papers to support their claim that EOM surgery is "controversial," you, the patient, should acquaint them with the facts, as documented in the referenced material.

Physicians who have understood and applied the latest scientific findings are using the above EOM surgeries. They are practicing the finest medicine, the kind that you and all other patients deserve; it is simply untrue to describe that as "aggressive." Conversely, the failure to offer patients the results of today's best science is not, in my opinion, good medicine. As both a potential patient and a recognized authority in the study of nystagmus, I would accept no less than the best available INS therapies for anyone in my care.

**REFERENCES**


1. Shallo-Hoffmann JA, Visco Jr. F, Tusa RJ. Mis-use of the artificial divergence operation to treat congenital nystagmus in a patient with infantile strabismus and acromatopsia: analysis of eye movement recordings. In: Sharpe JA, ed *Neuro-ophthalmology at the Beginning of the New Millennium*. Englewood: Medimond Medical Publications, 2000; 125-9.

2. Dell'Osso LF, Flynn JT. Congenital nystagmus surgery: a quantitative evaluation of the effects. *Arch Ophthalmol* 1979; 97:462-9.

3. Dell'Osso LF, Jacobs JB. An expanded nystagmus acuity function: intra- and intersubject prediction of best-corrected visual acuity. *Doc Ophthalmol* 2002; 104:249-76.

4. Anderson JR. The treatment of congenital nystagmus. In: *Ocular vertical deviations and the treatment of nystagmus*. Philadelphia: J.B. Lippincott Co, 1959; 1654-172.

5. Dell'Osso LF, Hertle RW, Williams RW, Jacobs JB. A new surgery for congenital nystagmus: effects of tenotomy on an achiasmatic canine and the role of extraocular proprioception. *J AAPOS* 1999; 3:166-82.

6. Hertle RW, Dell'Osso LF, FitzGibbon EJ, Thompson D, Yang D, Mellow SD. Horizontal rectus tenotomy in patients with congenital nystagmus. Results in 10 adults. *Ophthalmology* 2003; 110:2097-105.

7. Hertle RW, Dell'Osso LF, FitzGibbon EJ, Yang D, Mellow SD. Horizontal rectus muscle tenotomy in patients with infantile nystagmus syndrome: a pilot study. *J AAPOS* 2004; 8:539-48.

8. Wang ZI, Dell'Osso LF. Being "slow to see" is a dynamic visual function consequence of infantile nystagmus syndrome: Model predictions and patient data identify stimulus timing as its cause. *Vision Res* 2007; 47(11):1550-60.

9. Wang ZI, Dell'Osso LF. Tenotomy procedure alleviates the "slow to see" phenomenon in infantile nystagmus syndrome: model prediction and patient data. *Vision Res* 2008; 48:1409-19.

10. Bietti GB. Note di tecnica chirurgica oftalmologica. *Boll d'oculist* 1956; 35:642-56.

11. Bietti GB, Bagolini B. Traitement médico-chirurgical du nystagmus. *L'Annee Ther Clin Ophtalmol* 1960; 11:268-93.

12. Von Noorden GK, Sprunger DT. Large rectus muscle recession for the treatment of congenital nystagmus. *Arch Ophthalmol* 1991; 109:221-4.

13. Sprunger DT, Fahad B, Helveston EM. Recognition time after four muscle recession for nystagmus. *Amer Orthoptic J* 1997; 47:122-5.

14. Wang Z, Dell'Osso LF, Zhang Z, Leigh RJ, Jacobs JB. Tenotomy does not affect saccadic velocities: Support for the "small-signal" gain hypothesis. *Vision Res* 2006; 46:2259-67.

15. Tomsak RL, Dell'Osso LF, Rucker JC, Leigh RJ, Bienfang DC, Jacobs JB. Treatment of acquired pendular nystagmus from multiple sclerosis with eye muscle surgery followed by oral memantine. *DJO* 2005; 11(4):1-11.

16. Wang ZI, Dell'Osso LF, Tomsak RL, Jacobs JB. Combining recessions (nystagmus and strabismus) with tenotomy improved visual function and decreased oscillopsia and diplopia in acquired downbeat nystagmus and in horizontal infantile nystagmus syndrome. *JAAPOS* 2007; 11:135-41.

17. Wang ZI, Dell'Osso LF. A review of the tenotomy nystagmus surgery: origin, mechanism, and general efficacy. *Neuro-Ophthalmol* 2007; 31:157-65.

18. Dell'Osso LF. INOS 2008: ethical science or something else? OMLAB Editorial 2007; #071107:1-8. http://www.omlab.org/Editorial/editorial.html

19. Dell'Osso LF. Responding to more than a response: tenotomy improves INS waveforms. OMLAB Report 2007; #071107:1-12. http://www.omlab.org/Teaching/teaching.html

## *Citation*