

Does Eye Muscle Surgery Improve Vision in Patients with Infantile Nystagmus Syndrome (INS)?

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Clinical and electrophysiologic data collected for more than 50 years supports the novel hypothesis that eye muscle surgery improves vision and visual function (other than binocularity) in patients with infantile nystagmus syndrome (INS).¹ Infantile nystagmus syndrome, formerly called *congenital nystagmus*, occurs in approximately 1 in 600 to 1 in 5000 children.² Eye muscle operations for nystagmus can be classified into 9 types. The goals of the first 8 are to improve both the nystagmus and anomalous head or eye position, and include moving or removing parts of 1 to 3 muscles or tendons per eye. The ninth operation improves the nystagmus alone (removal of the 2 horizontal recti on each eye and reattachment at the original insertion).³ Eighty-five percent of the patients have procedures 1 thru 8, whereas only 15% of our patients have the 9th procedure).¹⁻³

Dodge's introduction of eye movement recording analysis at the turn of the 20th century has influenced many areas of scientific discovery profoundly. Fifty years ago, Anderson (of the Anderson-Kestenbaum procedure) said that in children, "both the nystagmus and vision improved after eye muscle surgery for torticollis."⁴ This was elaborated further in the electrophysiologic observations and reports by Flynn and Dell'Osso in the late 1970s and early 1980s and by animal work and National Eye Institute-supported clinical trials performed in the mid to late 1990s.⁵⁻⁷ An affirmative answer to the question, "Does eye muscle surgery in patients with INS improve recognition of black letters on a white screen, or its measure, that is, Snellen, Early Treatment Diabetic Retinopathy Study or AQ:1 HOTV acuity?" is important to clinicians and their patients. Unfortunately, high spatial frequency vision was, is, and continues to be a poor primary measure of ocular motor function. For example, a patient with INS and a latent AQ:2 nystagmus [right?] component has multiple optotype measures of acuity depending on mental state, attention, and whether fixation is monocular, binocular, at distance or near, or at eccentric gaze angles. Another 17% to 33% of patients with INS have a periodically changing cycle in which their null zone and best visual function exist not only at a place in space, but also during a period in time, making measures of acuity not only statically but also dynamically dependent.⁸ In patients with INS, measuring best-corrected binocular vision in and during the null zone is a visually unique ocular motor variable because of the nature of the oscillation.

Multiple examples of level II and III evidence suggest that eye muscle surgery improves nystagmus and visual function in patients with INS.^{1,9-12} Vision, used as an outcome measure when reporting effects of eye muscle surgery, must be explicit, because there are at least 5 psychophysical measures of acuity (i.e., detection, recognition,

resolution, hyperacuity, and localization). Variables reported before and after surgery in patients with INS include: optotype and gaze-dependent acuity, contrast sensitivity, motion detection, null zone characteristics, visual recognition time, subjective visual function, and electrophysiologic characteristics.^{1,7,10-12} In 138 patients from multiple studies who had their null zone optotype best-corrected binocular vision tested within 1 week and 4 to 6 weeks after eye muscle surgery for INS, grouped mean data showed a significant improvement ($P < 0.05$), and overall, 75% improved by 1 to 3 lines and 15% improved by 3 lines or more.^{7,10,13} Eye movement recordings also show that surgical intervention increases the prevalence of favorable nystagmus waveforms.^{7,10,13} Even if the nystagmus were eliminated completely, the integrity of the afferent system (optic nerve, retina, brain disease) and the age-related timing of surgery limit the acuity potential in any one patient with INS.

As a result of eye muscle surgery improving the beat-to-beat nystagmus, these patients receive more useful vision per unit of time and as a function of gaze, recognize objects faster and have less head movement and better motion and contrast sensitivity, and thus function better.^{7,10,13} The common clinical perception is that eye muscle surgery only serves to centralize the INS null position. In fact, what happens is a broadening and deepening of the null zone.¹³ In a more scientifically sound representation of this phenomenon, we have labeled this area *visual function space*, a 3-dimensional representation of visual function.

Data accumulated over the last 30 years show that many afferent and efferent visual system measures improve as a result of eye muscle surgery on INS patients, regardless of the indication (eccentric null, vergence damping, strabismus, or nystagmus alone), suggesting that neurovisual changes take place as a result of the surgical procedure itself, unrelated to moving or removing some of the extraocular muscle.^{1,7,10-12} The current hypothesis is that in surgical interference with peripheral extraocular muscle or tendon, enthesial proprioceptive nerve endings influence central ocular motor pathways, resulting in an improved INS oscillation.^{1,7,10-12} Anderson's observations of 50 years ago have been expanded. Research in nystagmus has begun to answer the question of whether eye muscle surgery improves nystagmus and visual function in the affirmative, but a multitude of questions remain.

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