# Extraocular Muscle Tenotomy, Dissection, and Suture: A Hypothetical Therapy for Congenital Nystagmus

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## INTRODUCTION

In 1979, the effects of the Anderson-Kestenbaum[1-2] procedure on the characteristics of congenital nystagmus (CN) were studied.[3-4] This surgery was designed to rotate the null region of a patient's CN to primary position and eliminate the preferred head turn seen in patients with non-primary-position null angles. Our studies demonstrated that, in addition to rotation of the CN null to primary position, the nystagmus null region was broadened and the nystagmus at gaze angles away from the null was also damped. Subsequent studies demonstrated that these effects were permanent and that the postoperative nulls did not return to intermediate gaze positions between the primary and preoperative null angles.[5-6]

Calculations made from the Figures in the original study[3] show that the preoperative null regions, extending 3° to 9°, were significantly broadened to postoperative nulls extending 23° to 35°. Similarly, the CN intensities (amplitude×frequency) 20° lateral to the postoperative nulls were reduced and ranged from 3.6% to 50% of their respective preoperative values.

Primary-position visual acuities improved from preoperative values of 20/60 to 20/100 to postoperative values of 20/25 to 20/40. Thus, the net result of the Anderson-Kestenbaum procedure (in addition to rotating the null to primary position) was to damp CN at all gaze angles and to enable improved visual acuity regardless of the position of a visual target.

The postulated mechanisms for the initially observed secondary effects included nonlinear changes in the ocular motor plant dynamics (ie, changes in the characteristics of extraocular muscle, tendons, Tenon's capsule, fatty and scar tissue interactions).[3] Subsequent damping of the CN during the first year was attributed to reduction of fixation attempt, lowering of anxieties and, possibly, scarring at the sites of the new muscle insertions.

Another possible mechanism involves changes to the recently described pulley system associated with extraocular muscles.[7] Although the exact mechanisms involved in these secondary effects were not known, it was thought from the time they were first recognized that they could be exploited therapeutically.

The generalized damping of CN would benefit patients with no convergence null who belong to any of the following groups:

- they have a null at or near primary position;
- they have no appreciable null at all; or
- they have no stationary null, because of aperiodic alternating CN.

Patients in any of these groups should not have a preferred head posture other than primary position. The CN damping produced by such a procedure would result from the measurable and permanent secondary effects (other

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than null rotation) of the original surgeries. Because the intensity of CN is also directly related to "effort to see" or "fixation attempt," reducing this effort by surgically damping the CN has a further damping effect on the nystagmus.

To perform the surgery designed to produce a generalized damping of CN, one should follow exactly the same surgical protocol used for the Anderson-Kestenbaum procedure, with the sole exception of the final resections and recessions. Instead, a tenotomy of each horizontal rectus is performed, with dissection of the perimuscular fascia and reattachment of the muscle to its original insertion.

For a patient with a near primary-position null, the postoperative CN should be damped in primary position, the null region should extend out farther from primary position, and the off-null CN should also be damped from its preoperative intensities.

For patients with no appreciable null or with aperiodic alternating CN, the postoperative CN intensities should be lower than preoperatively at all gaze angles. This procedure is muscle sparing and, if a particular patient does not experience significant damping, maximal horizontal recti recessions could follow; the latter is probably best left as the surgery of last resort.

The idea of damping CN by maximal recession of all horizontal recti[8] has always caused concern.[9] This is especially so in nonstrabismic patients who might suffer diplopia in extreme lateral gaze as a result of muscle weakness. Also, weakening the effectiveness of the muscles results in opposing central adaptive changes aimed at increasing innervation to these "weak" muscles. This plasticity is an inherent part of the ocular motor system and produces well-known effects in acquired weakness because of myasthenia gravis,[10-11] Lambert-Eaton syndrome,[12] or peripheral nerve palsies.[13-14]

In CN, the adaptive changes would tend to increase the nystagmus over time. The final results of such weakening procedures depend on the relative amounts of initial weakening and resulting plastic increase in innervation.

The generalized damping produced by the Anderson-Kestenbaum procedure raises a question about the percentage of the total initial CN damping seen after a maximal recession procedure that is produced by the above secondary effects. "Initial" is referred to as the damping before adaptive changes act to increase innervation to the muscles. If such damping is significant, the maximal recessions, with their counterproductive adaptive response, may be unnecessary.

Obviously, more study is required to provide answers to these questions. In the interim, perhaps what has already been learned from ocular motility studies can be used directly. The tenotomy, dissect, and suture procedure may provide a means for positive therapeutic intervention in those patients whose lack of convergence or stationary, non-primary-position gaze-angle nulls preclude either the bimedial recession or the Anderson-Kestenbaum procedures.

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