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Congenital Nystagmus Surgery

A Quantitative Evaluation of the Effects

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• **Nystagmus intensities at various gaze angles were studied both preoperatively and postoperatively, using accurate ocular motility recordings, in three cases of congenital nystagmus. In addition to shifting the nystagmus null, the surgery broadened the null region and resulted in an overall reduction in nystagmus intensity at all gaze angles. Surgical rotation also resulted in improved visual acuity in all cases. The postoperative acuity at 0° was better than the preoperative acuity at both 0° and the patient's preferred gaze angle (ie, the preoperative null angle). This was true not only for the two patients who showed an improved preoperative acuity with their head turn but also for the patient whose preoperative acuity did not substantially improve with her preferred head turn. Eye movement recordings have made it possible to accurately determine the amount of surgery required and to predict acuity increases even when undetectable during the preoperative clinical examination.**

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Surgical treatment for congenital nystagmus (CN) was first advocated independently and almost simultaneously by Anderson[1] and Kestenbaum.[2] Although their rationales for the surgical approach differed, their resulting operations achieved the same ends, namely, displacing both eyes away from the zone of least nystagmus. Since their initial reports, others have appeared, confirming their observations and adding a number of other important ones.[3-17] Of particular importance to this report are those that have documented an improvement in visual acuity following nystagmus surgery. In those series reporting such improvement, 18 of 34 patients obtained improved acuity in primary position following CN surgery.[3-6,10-17] Significantly, two large series,[13,16] comprising some 107 patients, failed to find any improvement in acuity after CN surgery.

There now exists a substantial body of knowledge on the operative indications, routine methods of measurement of the head turn, quantitative amounts of surgery, effects to be expected, and modification of the treatment plan to encompass associated conditions such as strabismus and amblyopia. On the other hand, the precise location of the null zone[18] and the nystagmus "intensity"[19] (this term is defined as the product of the nystagmus frequency [F] in hertz and the amplitude [A] in degrees [Hz°]) at various gaze angles are quantitative measures of the severity of the nystagmus. The effect of surgery on these parameters and the resulting visual acuity have never been quantitatively evaluated.

By using quantitative eye-movement measurements to clearly define the characteristics of the CN in three patients who satisfied preliminary conditions, we have documented several beneficial effects of surgical treatment as well as establishing new, more precise criteria for both the surgery and the increased visual acuity that can occur in these patients.

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MATERIALS AND METHODS

Eye Movement Recordings

Eye movements were recorded, using an infrared reflection technique described previously.[19] The full system bandwidth was dc-100 Hz for both eye position and velocity signals; the latter were obtained by electronic differentiation of the former. Subjects were seated at the center of a 1.14-m radius arc that contained red light-emitting diode targets spaced at 5° intervals. The subject's head was held fixed by chin cup and neck brace. After calibrating the system, which was linear to ±20°, recordings were made as the subject viewed each target in turn throughout the ±30° range of the arc. Nystagmus amplitude (A) was measured in degrees peak-to-peak, frequency (F) was measured in hertz, and their product intensity (I = A × F), as defined above, was plotted over the range of gaze angles.

The choice of nonaccommodative targets at 1.14 m was deliberate. Our experience has shown that, to avoid the variables associated with anxiety, one must minimize the effort to see, which is responsible for the intensification of CN. At this distance the damping effects of convergence on CN are not great; in fact, damping usually becomes noticeable at reading distance or nearer. By depriving the subject of an identifiable (accommodative) target, we remove the anxieties associated with the identification process. In this way, we can make an accurate, uncontaminated measure of the variation of CN with gaze angle as different target lights are activated.

Surgery

In arriving at a determination of the amount of surgery to be done, a number of factors were taken into account. Among these were the preoperative quantitative determination of the null zone and our own observation of the patient's employment of his head turn in casual seeing as well as when stressing the system by determining the visual acuity both monocularly and binocularly at distance and near. Still another factor was the severity of the head turn observable in old photographs. This is important, because the patient may, due to peer pressure, be forced to abandon the head-turn position of best acuity and accept blurred vision in exchange for getting rid of the cosmetically unacceptable turn. In our experience the null zone does not change its location but, rather, the patient adapts a less-than-optimum head position for cosmetic reasons. In addition, we evaluated, as critically as possible, the patient's and/or parent's desire for cosmetic improvement. During the examination itself, one must pay particular attention to the presence or absence of binocular single vision, strabismus, amblyopia, and the possibility of a fundus lesion that might limit the visual acuity postoperatively.

In the presence of good visual acuity, a cosmetically unacceptable head turn, and binocular vision, we performed surgery necessary to move the null zone from its position in eccentric gaze to the primary position. This surgery was performed on both eyes. For example, if the neutral zone was 20° to the right, this would require a leftward rotation of each eye 20°. In the presence of strabismus, only the fixing eye was operated on, and the surgery done was performed independent of its effects on the nonfixing eye. In some circumstances, this can lead to worsening of the strabismus, its reversal, or creation of a pseudostrabismus. We do not hesitate to reoperate and recorrect in these patients should the primary surgery prove inadequate or create or worsen the strabismus.

Visual Acuity

This was measured with Snellen optotypes in a 6-m lane illuminated by 20 foot-candles. The acuity was determined, where possible, monocularly, binocularly, in the head-turn position, in the primary position, and opposite the head-turn position. Near visual acuity was determined on the Lebensohn chart in the same fashion. Particular attention was paid to the head position assumed by the patient during testing of the visual acuity. This is the position where the patient places his eyes for his visual system to be maximally at rest, and often it is under these circumstances that the maximum head turn of the patient is elicited. These acuity figures were recorded in the patient's chart and were unknown to the author who performed the preoperative and postoperative eye movement recordings and whose predictions of acuity increases that would result from surgery were based solely on an analysis of the eye movement recordings.

REPORT OF CASES

CASE 1.—A 15-year-old girl was noted to have poor visual acuity after birth. Bilateral massive hemorrhages were noted at 10 weeks of age, and these cleared completely by 3 months of age. At 8 weeks of age, CN and a variable esotropia were noted; this later became a full-time left esotropia with amblyopia. Repeated attempts to patch the right eye during childhood failed to improve the vision in the left eye.

On examination, a head turn to the right was present and her binocular visual acuity for distance was 20/70 in straight-ahead position and 20/60– in left gaze. Monocularly, it was 20/70 in left gaze. Her near visual acuity was 20/60 in the right eye. In her left eye, visual acuity was 20/200 and unimprovable. She had 25 PD of left esotropia by prism corneal light reflex at both distance and near. Results of retinoscopy were +1.00 +0.75 × 95 in the right eye and +3.00 +1.00 × 85 in the left eye; this did not substantially improve her acuity or head turn.

Quantitative Nystagmus Characteristics (Before Surgery).—The nystagmus was jerk right, which converted to jerk left only beyond 25° left gaze. Her CN waveforms were jerk (J), jerk with extended foveation (J_{EF}), pseudocycloid (PC), and pseudojerk (PJ).[20] There was a sharp null at 20° left gaze (Fig 1). Attempted convergence actually resulted in both eyes moving to the left; the resulting nulling of the nystagmus in the right eye was not a true convergence effect. Despite her strabismus, amblyopia, and suppression of her left eye, no manifest latent nystagmus was present.[21]

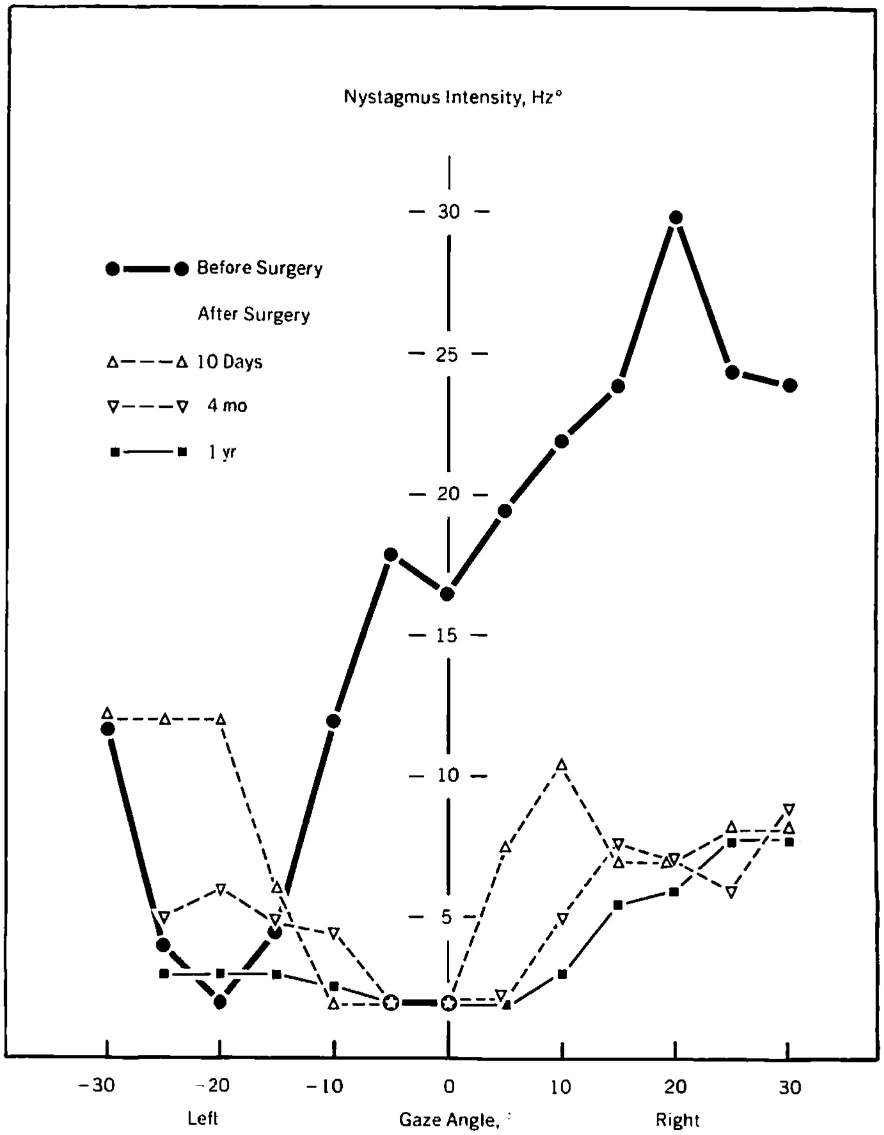

Fig 1.—Preoperative and postoperative plots of nystagmus intensity vs gaze angle for case 1, illustrating time course of improvement.

The finding of a sharp null at 20° left gaze indicated that, since the retina was normal, surgical rotation of the null to the straight ahead could still improve her acuity despite the lack of any major clinically determined acuity improvement in her head-turn position.

Surgery.—Surgery adequate for a 20° shift (40 PD) would be expected to overcorrect the 25-PD esotropia. The patient and her parents were informed of these considerations and elected to go ahead with the monocular surgery. The right medial rectus was recessed 5 mm, and the right lateral rectus was resected 6 mm.

Quantitative Nystagmus Characteristics (After Surgery).—The effects of surgical rotation on the nystagmus picture and the progression of these effects are shown in Fig 1. Not only was the null shifted but it was broadened and the nystagmus was much less intense surrounding the null than it was before surgery. During the first year after surgery there was a gradual improvement of these effects.

Course.—In the immediate postoperative period, the patient was 15 PD exotropic and her visual acuity was 20/70+1 in the primary position. On the 11th postoperative day, her visual acuity was 20/40-2. Her exotropia has remained unchanged. Her chosen gaze position was 5° to the left of the primary position, and she was given a prism of 9 PD base out over the right eye to enable her to use this position without a head turn. In this way prisms were used to "fine tune" the surgical results.[19-22] Postop-

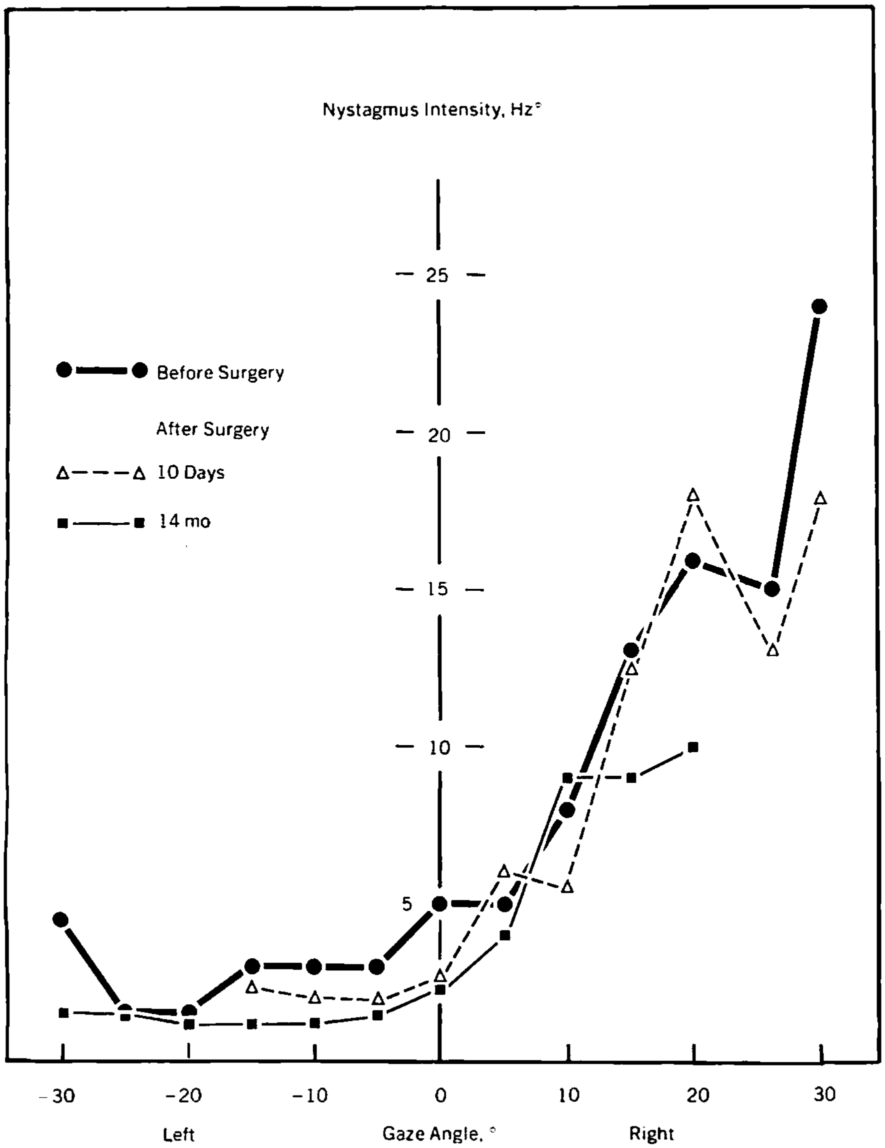

Fig 2.—Preoperative and postoperative plots of nystagmus intensity vs gaze angle for case 2, indicating time course of improvement.

erative acuity in the primary position improved to 20/40− binocularly, reflecting the factor of eight decrease in the nystagmus intensity.

CASE 2.—A 6-year-old boy had CN first noted at 2 or 3 months of age. A head turn to the right was present for an undetermined length of time. Results of detailed ophthalmologic examination were normal. The patient fused, had normal fusional amplitudes, and normal retinal correspondence on the synoptophore. With his head in the preferred position to the right (gaze left), his acuity was 20/50 in the right eye, 20/40 in the left eye, and 20/40+ in both eyes. When turned to the left, it was 20/200−. His near visual acuity was 20/30 binocularly at 20 cm.

Quantitative Nystagmus Characteristics (Before Surgery).—The nystagmus was jerk right except in far left gaze where it was jerk left. His CN waveforms were J, J_{EF}, PC, and bidirectional jerk (BDJ). There was a null between 20° and 25° left gaze as shown in Fig 2. Convergence also decreased his nystagmus intensity, but his right eye tended to drift in an outward direction. The presence of the null in left gaze (at approximately 22.5°) along with a normal retina indicated that surgical rotation would improve acuity in the straight-ahead position. In this case, the clinical evidence of improved acuity with a head turn supported the data from the eye movement records.

Surgery.—In contrast to the preceding patient, this patient presented the classical indications for the Anderson-Kestenbaum relocating operation.[11] The task here was to move both eyes at least 20° to the right. To do so, we chose the "5-6-7-8 operation," popularized by Parks.[3] Postoperatively, the patient immediately complained of diplopia and required varying amounts of prism from 4 to 12 PD base out over the right eye to fuse. His head turn partially recurred in the amount of 10° to the right. He continued to require prisms of up to 12 PD to fuse.

Quantitative Nystagmus Characteristics (After Surgery).—The nystagmus characteristics measured postoperatively showed a null shift toward the straight-ahead, a broadening of the null region, and a lower nystagmus intensity over the range of gaze angles tested (Fig 2). These effects continued during the first year, as can be seen by the data taken 14 months after surgery.

Course.—The problems presented by this boy postoperatively were twofold. First, he had a persistent surgically created esotropia of 12 to 16 PD, requiring prisms to fuse. Despite this, his acuity increased to 20/30. Second, his head turn partially recurred. It was therefore decided to selectively weaken his left turners, that is, his right medial rectus and left lateral rectus, doing proportionately more on the right medial rectus than on the left lateral rectus to overcome the esotropia. This was accomplished by a further 2-mm recession of the medial rectus (a total recession of 7 mm) combined with a 3-mm recession of the left lateral rectus (total recession of 10 mm). Esotropia was eliminated as a result of the second operation and the effect on the nystagmus intensity, 4½ months after surgery, was a broad range of low-intensity nystagmus centered around the primary position (Fig 3). Reflecting this improvement was a further acuity increase to 20/25.

CASE 3.—A 23-year-old man was born prematurely and developed retrolental fibroplasia (RLF). Nystagmus appeared in early infancy, and an esotropia for which he had surgery on the right eye appeared at age 8. He never had good vision in the right eye and was aware that he had to turn his head to the left to see clearly with his left eye. Examination revealed a vision of light perception with projection in the right eye; in the left eye his vision was 20/100 in the primary position and 20/60 in right gaze. At near, he saw 20/30 with the left eye. He had 20 PD of a right esotropia by prism corneal light reflex method. A poor direct light reflex and an afferent pupillary defect were present on the right. Slit-lamp examination showed early band keratopathy, a retrolental membrane, and a peripheral traction detachment in the right eye. In the left eye, there was marked straightening of the vessels, ectopia of the macula, pigment changes, and traction in the periphery consistent with a diagnosis of regressed RLF (Fig 4).

Quantitative Nystagmus Characteristics (Before Surgery).—The nystagmus was totally jerk left within the ±30° range of gaze angles tested and decreased toward a null that was beyond 30° right gaze (Fig 5). His CN waveforms were J, J_{EF}, and PC. The nystagmus intensity in left gaze was very high, making vision in that direction extremely difficult. Attempted convergence also nulled the nystagmus. No manifest latent nystagmus was found.[21]

Surgery.—This patient presented the typical indications for an immobilizing type of operation,[11] ie, a monocular patient with a null position in far eccentric gaze (30° to the right). Surgical rotation here, which must be accomplished on the fixing eye, would be of such magnitude as to clearly overcorrect his esotropia (20 PD). In addition, the heterotopia of the macula adds to the real divergence that might occur postoperatively, a pseudodivergence due to his large positive angle kappa. Nevertheless, because of the severity of the cosmetic defect and the improvement of the visual acuity in the head-turn position, the patient decided to go ahead with surgery. A recession of the left medial rectus of 8 mm and a resection of the left lateral rectus of 7 mm were performed.

Quantitative Nystagmus Characteristics (After Surgery).—As is evident in Fig 5, a dramatic decrease in nystagmus intensity resulted from the surgery. The null extended throughout the right field of gaze, and much of the left field was recovered for useful vision, as is shown by the data taken nine months after surgery. The intensity at straight-ahead was reduced by a factor of ten.

Course.—Postoperatively, the patient appeared exotropic due to the heterotopia but, significantly, had no true exotropia. The nystagmus intensity reduction was reflected by the acuity, which improved to 20/30−1 in the primary position. A 12 PD base-out prism was recommended for the left eye to "fine tune" the surgical results, but the patient was lost to further follow-up.

COMMENT

Surgical Indications

From the standpoint of the physician faced with a child with CN and a disfiguring head turn, a number of clinical points should be kept in mind. Surgery is best performed in the preschool years to alleviate the cosmetic defect prior to this very sensitive psychological period. In some cases this may preclude the type of recordings that are considered in this article. Second, cosmetic relief of the head turn can be accomplished in most cases, employing the principles formulated by Anderson[1] and Kestenbaum[2] with the modifications added by other clinicians during the past 25 years. In the presence of good binocular vision and ocular alignment, surgical rotations are accomplished by weakening the yokes that turn the eyes toward the null position and strengthening the antagonists, thereby rotating the eyes in the opposite direction. In the presence of strabismus and amblyopia (as is often the case in these patients), surgery is restricted to the fixing eye only.[7] When the deviation is esotropia and the null zone is in adduction, surgery has a beneficial effect on both, provided the surgical rotation

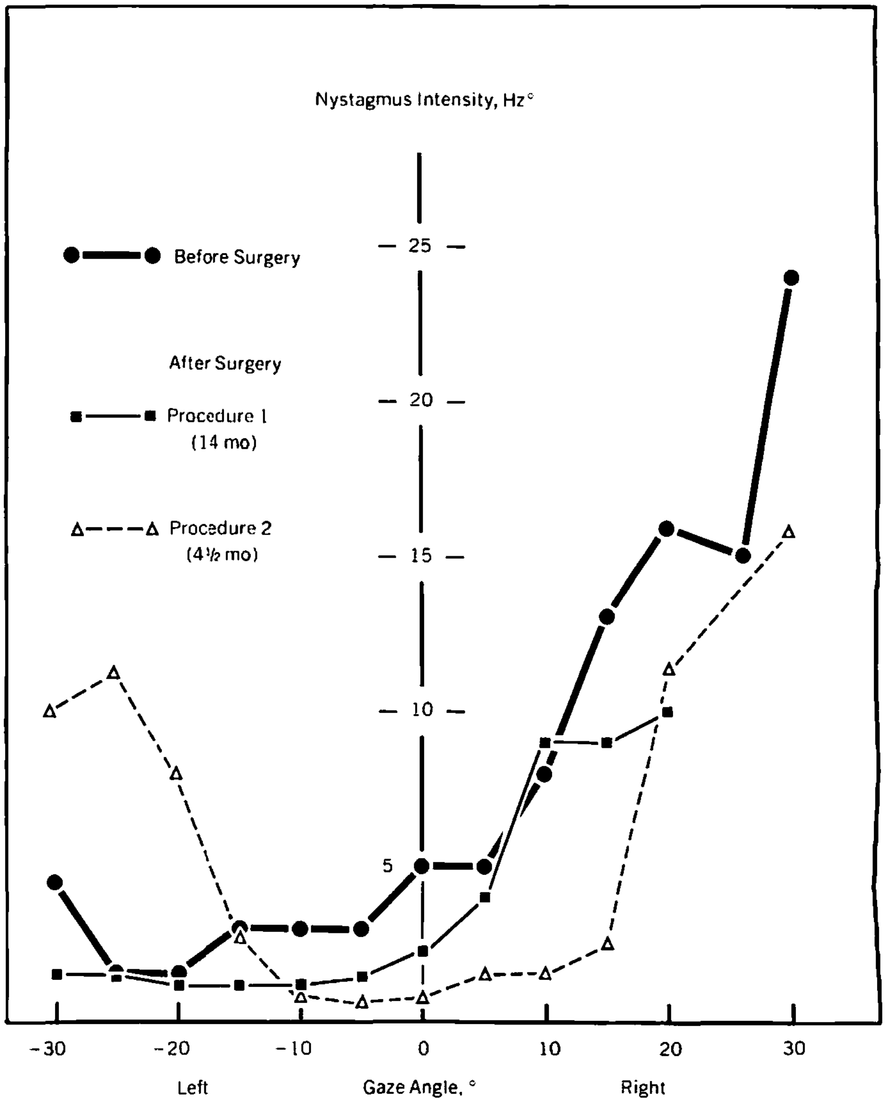

Fig 3.—Preoperative and postoperative (procedures 1 and 2) plots of nystagmus intensity vs gaze angle for case 2.

Fig 4.—Fundus of left eye of patient 3, showing straightening of vessels and ectopia of macula.

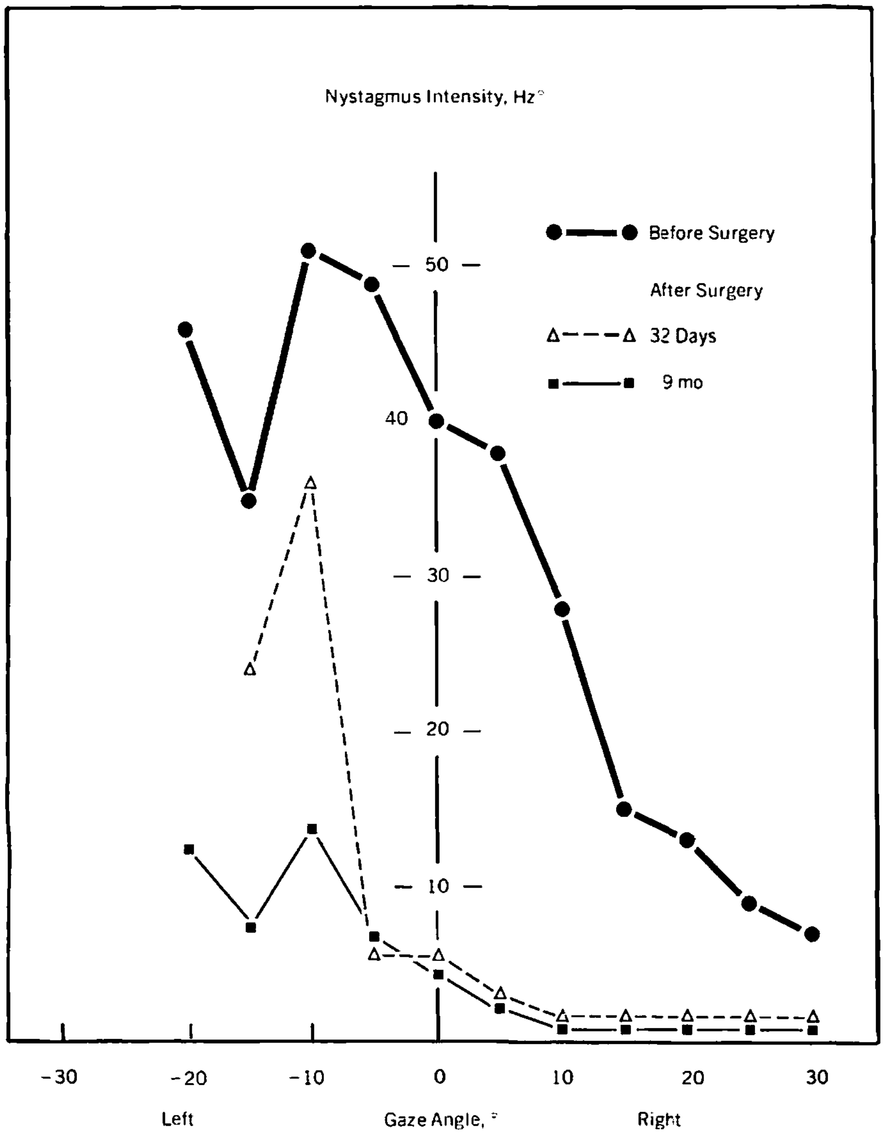

Fig 5.—Preoperative and postoperative plots of nystagmus intensity vs gaze angle for case 3, indicating time course of improvement.

required is not much greater than the size of the tropia. In the presence of esotropia with the null zone in abduction of the fixing eye, surgery to shift the null zone inevitably aggravates the cosmetic appearance of the turn. We believe this must be corrected at a second procedure, although other opinions exist on this point.[7] The opposite is the case with exotropia. If the fixing eye is abducted, surgery will both shift the null zone toward the primary position and improve the exotropia. If the fixing eye is in adduction, surgery, while it shifts the null toward the primary position, will aggravate the exotropia. Fortunately, in our experience, the first case—esotropia with the null zone in adduction—is the most common presentation of strabismus with congenital nystagmus.

In addition to the foregoing, when one is seeking possible improvement in the visual acuity by means of CN surgery, a very careful evaluation of the fundus for acuity-limiting defects in the macula or fovea is essential. As determined by case 3, however, visual improvement can occur even in the presence of macular damage from acquired disease. This is an exceptional case, and the surgeon would be well advised to be guarded about possible acuity effects in the presence of retinal lesions. As has been pointed out, if the patient exhibits a sharply defined null on quantitative recordings, this suggests that the acuity may improve with an operation. This is true even when, on clinical visual acuity testing, no improvement of the acuity is discernible in the head-turn position. Case 1 made the initial and most important contribution in establishing this new, broader criterion for surgical intervention. The patient had a sharply defined null and no fundus lesion that would in any way limit her acuity if the eyes could be still. What set her case apart was the lack of any substantial clinical improvement in the acuity before surgery in her head-turn position. Before quantitative recordings, given such a clinical finding, no acuity increase would be expected. In fact, the patient and her family were told that only a cosmetic improvement could be expected from surgery, but a study of her recordings and our experience with prisms indicated that we should expect a major acuity improvement. The results of her surgery confirmed the prediction, which was based on the quantitative recordings, and in so doing expanded the diagnostic criteria for surgical therapy of CN to include the possibility of improved acuity as a reason for surgery. Case 2 also provided strong evidence for the importance of surgical rotation in increasing visual acuity. Following the first surgical procedure, the acuity was substantially higher than the preoperative acuity. This acuity improvement was maintained in spite of the presence of a surgically created esotropia and diplopia, which required further correction (see below).

Effects of Surgery as Documented by Quantitative Recordings

There are three major beneficial effects of surgery that can be documented by our recording techniques: the desired null shift, broadening of the null zone, and the overall reduction of nystagmus intensity at off-null gaze angles. For purposes of discussion of the surgical effects of null broadening and overall intensity lowering, we will employ the term "usable null shift." The usable null shift is defined as the difference between the preoperative null and the most central position of the postoperative null region in which the nystagmus intensity is within 1 Hz° of its absolute minimum. The definition also yielded the actual usable null region for the patient, which is defined as the region of gaze lying within 1 Hz° either side of the absolute null. The Table lists these values for each subject, along with the preoperative and postoperative acuities in the primary position at distance.

This quantitative study of surgery for CN has uncovered effects of the surgery not immediately apparent to the clinical observer. In addition to the expected null shift as a result of surgery, we found two other changes in the nystagmus intensity plots that might be considered conducive to better acuity. The first was a broadening of the null region, which provided a wider range of gaze angles with minimal nystagmus. This made the actual gaze angle employed by the subject less critical and thus good acuity easier to maintain. The second was the overall reduction of the nystagmus intensity outside the null zone. With it comes a corresponding reduction of effort to see on the part of the subject. This effort to see involves the subject with nystagmus in a vicious cycle of more effort intensifying his nystagmus, making it more difficult for him to see.[23] In any case, we speculate that this improving nystagmus intensity with time may be secondary to a general lowering of the patient's anxiety level as they learn that good vision is now present at a glance without the effort that had been previously required to accurately find and hold the narrow range of gaze angles where the nystagmus intensity was minimal.

The net effect is the relationship between the reduction of the nystagmus intensity and the improvement in visual acuity. Figure 6 is a plot of data derived from all three cases. In each case, the nystagmus intensity and visual acuity in the primary position are indicated by the open circles preoperatively and the filled circles postoperatively. In all three cases, a substantial reduction of the nystagmus intensity in the primary position has occurred as a result of the surgery. The letter N indicates the nystagmus intensity and visual acuity at the patient's chosen null position before surgery. The difference between the nystagmus intensity at the null position before surgery and the primary position after surgery is not striking, yet there is an improvement in visual acuity. Why? Consider the data from case 1. If a sharply defined null existed, why then was the acuity not better with the head turned? Why was the postoperative acuity at 0° better than the preoperative acuity at the null position? The nystagmus intensity for these two positions of gaze was virtually the same. We believe the explanation for this apparent paradox becomes clear when one considers the conditions under which the data for Fig 6 were obtained. The acuity data, both before and after surgery, were obtained in the clinical setting with the patient involved in performing a task that gets more difficult as his actual maximum acuity level is approached. Thus, since the patient is increasing his effort to see (fixation attempt) during the actual assessment of his acuity, he can never reach the level of acuity that is his potential. If a way could be found to prevent or diminish this increasing effort to see, he might well reach his potential. The intensity data of Fig 6, on the other hand, were gathered in such a way as to minimize his effort to see. All target lights in the arc were equally and easily visible to the subject and unlikely to provoke increasing effort to see. The variation of the nystagmus intensity with gaze angles at this low level of anxiety-effort is a more sensitive indicator of possible acuity improvement, given that the precondition of no acuity-limiting ocular condition is present. Thus, the apparent contradictions of Fig 6 are actually the results of different types of data gathered under very different conditions. In those patients who do have a preferred head turn, this is prima facie evidence that their nystagmus is minimal at that gaze angle and that their acuity is potentially better there (whether this can be measured clinically or not). Ocular motility recordings accurately document these cases and indicate those patients whose nulls are sharp enough to allow for improved acuity. Case 1 is the prime example of this. Case 2 seems to present a contradiction in that the postoperative nystagmus intensity at the primary position after the first procedure is slightly greater than at the preoperative null, yet the acuity is higher at the primary position. This, we believe, is an artifact of the different conditions under which these two cases of data were collected. Consider, finally, the data for case 3, which we classify as CN/acquired.[21-24] In this special case, we have the unique opportunity to study a nystagmus that we can safely presume was acquired in early infancy (secondary to early-acquired disease of the retina). In the majority of cases, one cannot separate a true CN from a CN/acquired, since early documentation of the absence of nystagmus at birth is not available. Thus, the designation "CN" is retained with a "/A" designation added when such documentation is available. It is noteworthy that in this documented case of early-acquired nystagmus the waveforms were jerk (J, J_{EF}, PC) and not

Effects of Congenital Nystagmus Surgery on Null and Visual Acuity

Subject	Usable Null Shift, °	Usable Null Region, ° Before Surgery	Usable Null Region, ° After Surgery	Visual Acuity (0°) Before Surgery	Visual Acuity (0°) After Surgery
1	20	3.5	35	20/60	20/40
2	21.5 (22.5)*	9	29 (17)*	20/60 +	20/30 (20/25)*
3	22.5	3	23	20/100	20/30

*These figures are the results of the second operation.

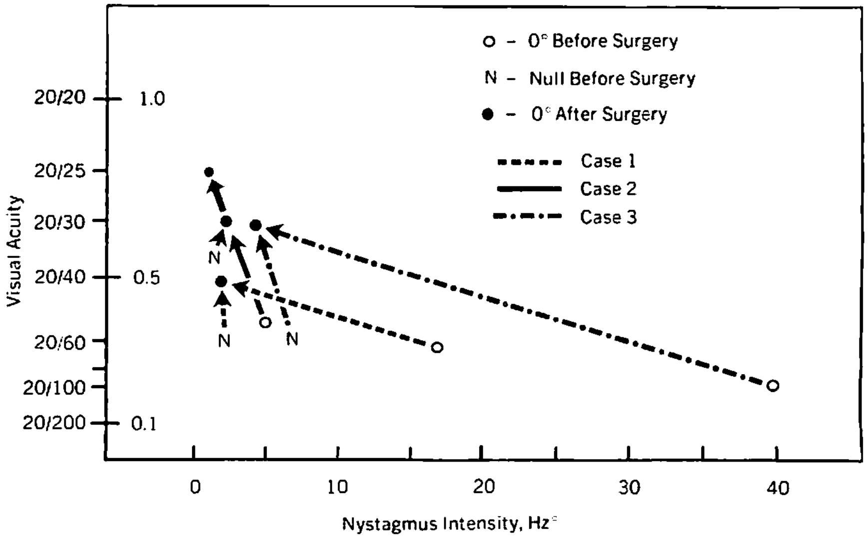

Fig 6.—Preoperative and postoperative visual acuities and nystagmus intensities for patients 1, 2, and 3, including both postoperative points for patient 2.

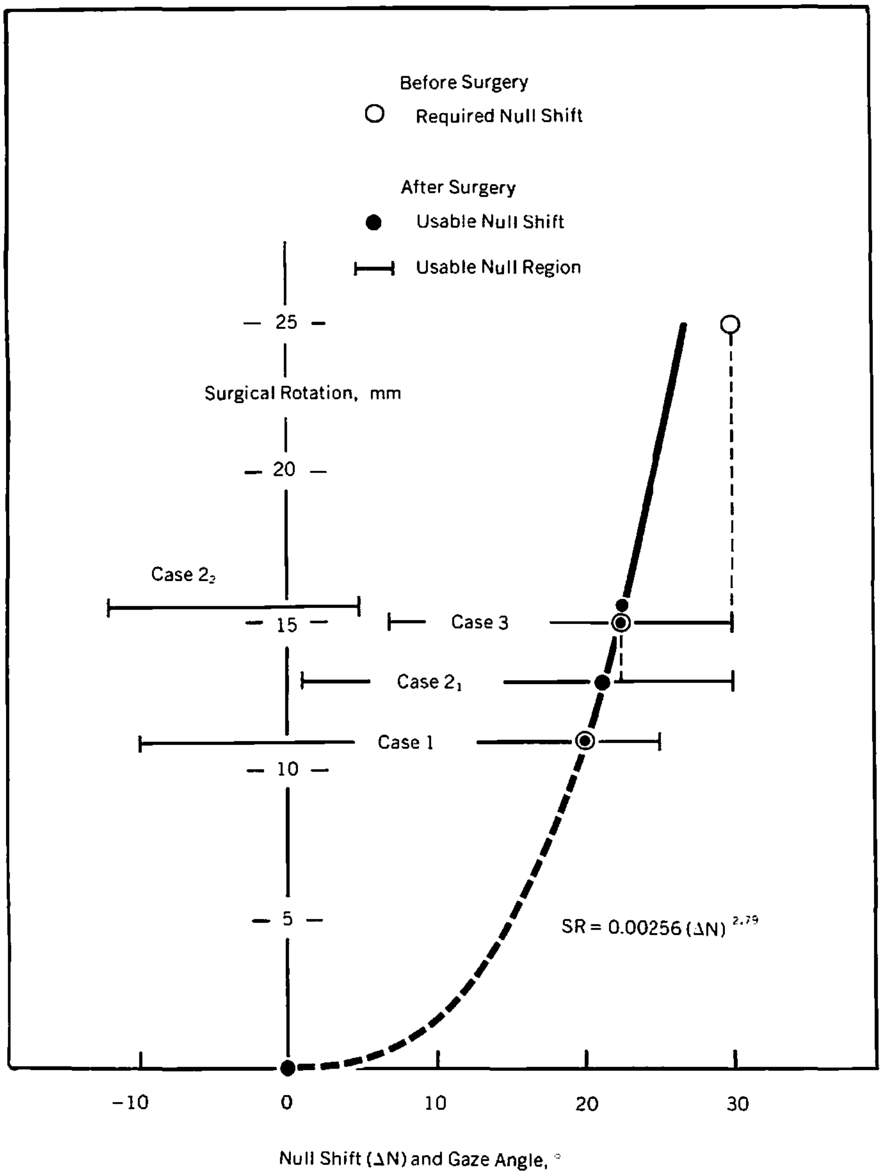

Fig 7.—Relationship between total surgical rotation and resulting usable null shift in patients 1, 2, and 3, including both operations for patient 2, is shown with accompanying usable null regions. All data are plotted in same direction for clarity; actual null shift directions are shown in Fig 1 through 3 and 5.

pendular. At present these two forms of nystagmus, CN and CN/A, cannot be separated either clinically or by means of quantitative recordings. Patient 3's waveforms, which reflect early developmental changes designed to increase the foveation time per cycle, are the same and differ from those of latent nystagmus[21] and various other forms of nystagmus acquired later in life. We see from Fig 6 that the postoperative acuity at 0° was greater than the preoperative acuity at the null, and that the intensity was also less after surgery. Suffice it to say, in summary, that given the preconditions for intervention outlined under surgical indications, we can anticipate acuity at the primary position to be at least as good, and probably better than, at the previous null angle.

It is not immediately obvious why a simple globe rotation should result in both a broadening of the null and an overall reduction in nystagmus intensity and how these two effects can, in turn, be linked to clinical improvement in the patient's acuity. Inspection of the illustrations, however, shows that both effects become more pronounced with time, especially the overall reduction of nystagmus intensity. It is possible that the immediate postoperative broadening is due to nonlinear changes in ocular motor plant dynamics (ie, changes in the characteristics of the muscles, tendons, Tenon's capsule, fatty and scar tissue interactions) as a result of the surgical changing of the points of insertion and methods of attachment of the muscles to the globe. The further reduction of the nystagmus during the first year after surgery (which tends to further broaden the null zone) is probably due to the lowering of both the patient's fixation attempt on testing and the psychological anxieties historically associated with his previous attempts to see. Additional factors may be further dynamic changes due to scarring at the new muscle insertions. In addition, we cannot completely discount the possibility of a training effect occurring in these subjects on repeated recording of their nystagmus. It is noteworthy, however, that repeated recording for many years of the nystagmus of one of us (L.F.D.) produced no changes in the nystagmus intensity. We have also considered the possibility that the improvement of visual acuity was in fact a training effect in which the improvement occurred due to the patient's repeated practice. However, when we compiled our acuity data, we found no such trend in the measured acuities in any of the cases.

Surgical Rotation and the Resulting Null Shifts

To arrive at an accurate assessment of what surgery was accomplishing, certain simplifying assumptions were made to permit quantitative treatment of the amounts of surgery done. The millimeters of surgery performed on an eye (recessions and resections) were simply added to produce a number reflecting the total millimeters of rotation of the eye. In Fig 7 the filled circles form the plot (for one surgeon) of the total surgical rotation (SR), in millimeters, vs the usable null shift obtained (ΔN), in degrees. We then compared these points with the original preoperative required null shifts, which are drawn on the above curve (open circles). The surgical data are described by the equation $SR = 0.00256(\Delta N)^{2.79}$. This curve is a "best fit" ($r^2 = .98$) to the actual surgical data of the initial operations on the three patients. The origin is shown as a data point, since zero surgical rotation would not cause any null shift. The dashed region is one for which data are presently lacking. The validity of this curve was strengthened by the fact that the point generated by the second procedure on case 2 fell on the already-existing curve.

As can be seen from case 1, as long

as the null region encompasses the 0° gaze angle, the usable shift cannot exceed the required shift (ie, you cannot overshift and the two points are coincident). The extent to which the null-broadening effect of the surgery overcomes less than adequate surgical rotation can be seen by the degree to which the usable null region overlaps or approaches the primary position (0°). In case 2, with 13 mm of total surgical rotation, because of the persistent esotropia and recurrence of a minimal head turn, we were afforded the opportunity to further rotate both eyes and, at the same time, to correct the esotropia. The initial 7-mm recession of the left lateral rectus was augmented by 3 mm, for a total of 10 mm, and the initial 5-mm recession of the right medial rectus was increased by 2 mm, for a total of 7 mm. The net result of both surgical procedures was a 16-mm rightward rotation of the left eye and a 15-mm rightward rotation of the right eye. Since the rotations were not equal, the mean value (15.5 mm) was chosen as the data point to be plotted in the illustration. The resulting null shift was exactly the amount that was initially required (22.5°) and that was predicted by the curve. The new 17° usable null region now encompassed the primary position and extended from 5° left gaze to 12° right gaze, as can be seen from the illustration. Although the final usable null region of 17° was somewhat less than the null region present after the first operation, it is still substantially better than the 9° preoperative region. More important than its absolute magnitude, however, is its location with respect to the primary position. Since the final null region straddles the primary position, it is much more conducive to lowered nystagmus intensity and increased acuity in this position. Bahill et al[25] have pointed out that most saccades are less than 15° in amplitude, which means that the eyes are almost always at gaze angles near the primary position. Minimizing the nystagmus in the central 30° of gaze allows for good acuity at the gaze angles most utilized under normal conditions.

It must be emphasized that this curve was derived for one surgeon following three surgical procedures in an attempt to quantitate the effect of his surgery. It was not employed to make the decision as to how much surgery would be undertaken in a given case to achieve the desired rotation, even though the curve was available before the second procedure in case 2. Inspection of the data, however, suggests that once the required preconditions for CN surgery are met and the decision to operate is made, the time for conservatism is over and the full amount of surgical rotation required should be performed. If comparable data are available, such a curve could be used to convert the required rotation, as measured from the nystagmus intensity plot, to millimeters of surgical rotation. The null-broadening effect will help negate slight underrotation and, since the amount of broadening is variable and overrotation does not seem probable, full required rotation should be attempted in each case.

Fine Tuning With Prisms

The use of prisms to achieve nystagmus nulling and increased acuity in cases of CN where the null is close to primary position was the basis for this study.[19,22,23,26] Their use to "fine tune" the results of surgery has also been suggested in previous studies.[19,22] Prisms can be used to shift gaze to the null angle when the surgical procedure falls slightly short, as in case 3, or to utilize the preferred gaze angle that lies within the new null region, as in case 1. The key element in determining the exact angle which a patient with CN prefers (ie, the position of best acuity) is the "foveation time" achieved by the particular waveform used at that gaze angle.[19,27] Nystagmus intensity is a good indicator of foveation time, since, for any given CN waveform, the lower the intensity the greater the foveation time. However, in the region of the null there may be very slight changes in waveform at certain gaze angles that do not result in intensity changes but do increase foveation time. This was the reason that, despite an absolute null region extending from 5° left gaze through 0° to 5° right gaze, patient 1 chose to use a gaze angle of 5° to the left. The addition of 9 PD base out to her right eye allowed her to use this preferred gaze angle with her head straight.

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