

CONGENITAL NYSTAGMUS SURGERY

PROF. L. F. DELL'OSSO

By Invitation

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ABSTRACT

Anderson-Kestenbaum operations for congenital nystagmus with severe head turn have been employed for the past twenty-five years. They have been documented as being effective in correcting head turns associated with congenital nystagmus. We have studied a group of patients with congenital nystagmus and head turn, both pre- and post-operatively, by means of recordings of eye movements. We have documented the effect of surgery in shifting the null position, in broadening the null position and lowering the nystagmus intensity. In some cases, an improvement of the visual acuity has also occurred.

Anderson¹ and Kestenbaum² independently and almost simultaneously first advocated surgical treatment of congenital nystagmus (CN) with a head turn. Although their surgical approaches differed, the results of both of their operations achieved the same ends, namely, displacing the eyes away from the zone of least nystagmus. Since their initial reports, others have confirmed their observations and added a number of other important ones. Goto³, on the basis of his EOG recordings, advocated resection and advancement of the muscles responsible for the quick phase of the nystagmus and also noted an improvement in the visual acuity of three of his four patients so treated. Pierce⁴ extended the principles advocated by Anderson and Kestenbaum to vertical head tilts accompanying CN. He also noted improved visual acuity in one of his two patients following such surgery. Sternberg-Raab⁵ reported the results of surgery in ten cases. Two of the ten had improvement in visual acuity following the surgery. In three of the ten, however, the head turn recurred on prolonged follow-up. Cooper and Sandall⁶ initially reported seven patients whom they treated surgically. They described a method of quantitative evaluation of the degree necessary to turn the eyes using the arc perimeter and advocated conservatism in the surgical approach. Two of their patients had improvement in their visual acuity following surgery. In a later report⁷, they extended their observations to eighteen patients with strabismus as well as CN. Schlossman^{8,9} described his treatment of CN accompanied by strabismus and built his surgical approach around a maximum 5 mm weakening procedure on the medial rectus and no more

than 1 mm. more recession or resection on the lateral rectus. He noted improvement in visual acuity in the primary position in at least one of the six patients he presented. On the other hand, Taylor¹⁰ recommended large lateral rectus recessions (8-9 mm.) and medial rectus recessions (6 mm.) combined with 6 mm. resections of their antagonists to effectively prevent the recurrence of the head turn. He described the synoptophore as an instrument adequate to measure the amount of eye deviation the surgeon had to correct to shift the quiet zone of the nystagmus to the primary position. Both of the cases he reported had significant improvement in their visual acuity following surgery. Crone¹¹ described two types of operations for congenital nystagmus. A "relocating" operation which is the classical Anderson-Kestenbaum operation and an "immobilizing" operation, first described by Friede¹² in 1956, which consists of retroequatorial displacement of the agonist muscle(s) producing the slow phase of the nystagmus. Kommerell¹³ presented a series of twenty-eight patients operated on by the Anderson-Kestenbaum operation. Twelve of his patients had full binocular vision, sixteen had strabismus and amblyopia or microphthalmos of one eye. Considerable improvement in the head turn was obtained in eighteen, slight improvement in four and no improvement in six. Significantly, he obtained no improvement in the visual acuity in any of his patients. Parks¹⁴ described quantitatively his approach to the surgery of CN with a head turn consisting of a 7 mm. recession of the lateral rectus and a 5 mm. recession of the medial rectus muscles turning the eyes into the position of least nystagmus and a 6 mm. resection of the medial rectus and an 8 mm. resection of the lateral rectus turning the eyes in the opposite direction, the so-called "5-6-7-8" operation. However, Calhoun¹⁵ found, in a series of nineteen patients operated on for CN with a head turn, that these indications were inadequate. He recommended doing 40% more on each of the muscles operated on to prevent recurrence of the head turn. Huggonier¹⁶ presented his experience with seventy-nine cases of surgically treated CN. Twenty-three had pure CN, fifty-six were associated with strabismus, twenty of the twenty-three with pure CN had a "good" result from surgery, defined by the author as improvement in the head turn and acuity in the

primary position. Of the cases associated with strabismus, in only fifteen was the nystagmus considered the primary indication for surgery. Thirteen benefited by the author's non-specific criteria. Fells and Dulle¹⁷ reported improved visual acuity in eight of nine patients after surgical correction of the compensatory head posture.

In summary, there now exists a substantial body of knowledge on the operative indications, clinical methods of measurement of the head turn, amount of surgery and modification of the treatment to encompass strabismus and amblyopia, conditions often associated with CN. On the other hand, the precise location of the null zone¹⁸ and the nystagmus "intensity"¹⁹ (this term is defined as the product of the nystagmus frequency (F in Hertz) x the amplitude (A in Degrees) 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 more precise criteria for both the surgery and the increased visual acuity which can occur in these patients.

MATERIAL AND METHODS

Eye Movement Recordings

Eye movements were recorded using an infra-red reflection technique described previously¹⁹. 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 meter radius arc which 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) in Hertz and their product intensity (I = A x F) as above defined was plotted over the range of gaze angles.

The choice of non-accommodative 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 significant; 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 at 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 pre-operative quantitative determination of the null zone; 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, the possibility of a fundus lesion which might limit the visual acuity post-operatively.

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 of 20°. In the presence of strabismus, only the fixing eye was operated upon and the surgery done was performed independent of its effects on the non-fixing eye. In some circumstances, this can lead too worsening of the strabismus, its reversal or creation of a pseudo-strabismus. We do not hesitate to reoperate and recorrect these patients should the primary surgery prove inadequate, create or worsen the strabismus.

Visual Acuity

This was measured with Snellen Optotypes in a 20 foot 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 very 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 pre- and postoperative eye movement recordings and whose predictions of acuity increases which would result from surgery were based solely on an analysis of the eye movement recordings.

CASE REPORTS

Case Report 1. A fifteen year-old white female was first noted to have poor visual acuity soon after birth. Bilateral massive retinal hemorrhages were noted at ten weeks of age. These cleared completely by three months of age. At eight weeks of age, CN and a variable esotropia was 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 the patient's binocular visual acuity for distance was 20/70 in straight ahead gaze and 20/60 in left gaze. Monocularly, it was 20/70 in the right eye in gaze to the left. Her near visual acuity was 20/60 in the right eye. In the left eye, she was 20/200 unimprovable. Twenty-five prism diopters of left esotropia by corneal light reflex was present at both distance and near. Her retinoscopy was: O.D. + 1.00 + 0.75 x 95 and O.S. + 3.00 + 1.00 x 85. This did not significantly improve her visual acuity or head turn.

On quantitative nystagmus recordings preoperatively, there was a sharp null at 20° to the left (Fig. 1).

The management problems that this patient presented were as follows. First of all, she had strabismus with amblyopia in addition to her CN. Surgery, to be effective, had to be confined to the fixing right eye. The task was to move the eye 20° to the right. To accomplish this, any surgery done would have to overcorrect her 25 prism diopter esotropia. After explaining these considerations to the patient and her parents, they elected to go ahead with the surgery and on 5/6/74, a right medial rectus recession of 5 mm. combined with a right lateral rectus resection of 6 mm. was performed.

Postoperatively, the patient was 15 prism diopters exotropic and has remained so to the present time. In the immediate postoperative period, the binocular visual acuity was 20/70 + 1 in the primary position. On the eleventh postoperative day, her visual acuity binocularly had improved to 20/40-2 in the primary position. The patient adopted a gaze position 5° to the left of the primary position. For this, she was given a prism of 9 diopters base-out over the right eye to enable her to use this position without a significant

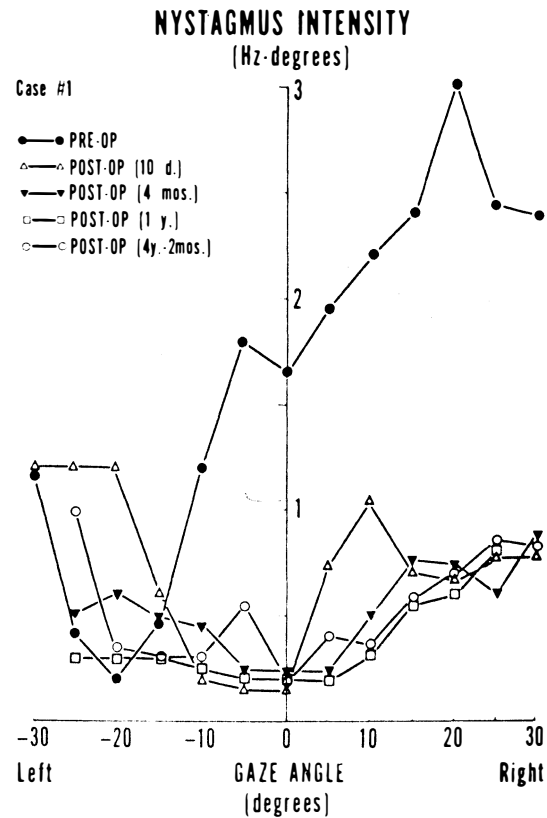


Fig. 1. Pre- and postoperative plots of the nystagmus intensity vs. gaze angle for Case 1, indicating the time course of improvement.

head turn. In this way, prisms were used to "fine tune" the surgical result^{19,20}. On last follow-up (7/11/78), the patient's binocular visual acuity remains 20/40-2 in the primary position at distance and 20/50- at near. Her postoperative recordings (Fig. 1) performed ten days, four months and one year and more than three years following surgery show reduction of the nystagmus intensity overall, shift in the null zone toward the primary position and a broadening of the null zone. In addition to this, the patient seems to have achieved a stable increase in her visual acuity.

To the clinician, this patient presented the problem of performing muscle surgery on the seeing eye for relief of the head turn. The second problem was the obvious necessity of performing surgery adequate to shift this fixing eye 20° to the right which would necessarily overcorrect the left esotropia. The risks attendant upon the performance of this surgery were acceptable to the patient and her parents. Postoperatively, we failed to achieve complete correction of the head position, but the patient has done very well since surgery with the use of a small prism before her fixing eye. She is now pursuing her career as a college student.

Case Report 2. A six year-old white male had congenital nystagmus with a right head turn, first noted at 2-3 months of age. Detailed ophthalmological examination was normal. When first seen by us in November, 1973, the patient's binocular visual acuity in his head turn position was 20/40, his monocular acuity was 20/50 in the right eye and 20/40 in the left eye. Out of his head turn position, it was 20/200-. His near visual acuity was 20/30 binocularly at 8 inches. The patient had fusion, normal fusional amplitudes and normal correspondence on the synoptophore.

On quantitative recordings of his nystagmus, it proved to be jerk right except in far left gaze where it was jerk left. There was a null between 20° and 25° left gaze preoperatively (Fig. 2). Attempted convergence increased his nystagmus.

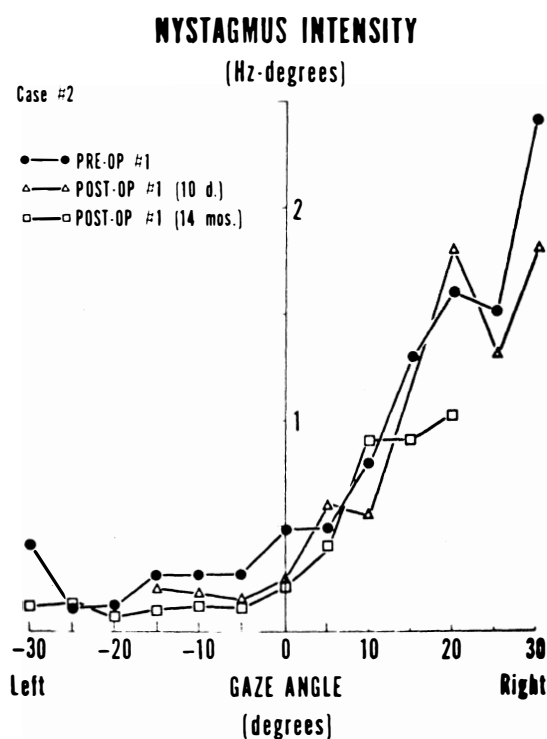


Fig. 2. Pre- and postoperative (procedure No. 1) plots of the nystagmus intensity vs. the gaze angle for Case 2.

In contrast to the previous patient, this patient presented the classical indication for the Anderson-Kestenbaum relocating operation. The task was to move both eyes 20° to the right. On 6/17/74, the left lateral rectus muscle was recessed 7 mm., the left medial rectus muscle was resected 6 mm., the right medial rectus muscle was recessed 5 mm., and the right lateral rectus muscle was resected 9 mm.

Postoperatively, the patient complained immediately of diplopia. He was found to have an

esotropia of 12-15 prism diopters. He fused with 12 prism diopters base-out before his right eye. The head turn partially recurred soon after surgery in the amount of 10° to the right. The nystagmus characteristics, measured postoperatively (Fig. 2), showed a null shift toward the primary position, a broadening of the null region and a lower nystagmus intensity over the range of gaze angles tested. These effects persisted through recordings up to fourteen months postoperatively.

The problems presented by this boy postoperatively were two-fold. First, he had a persistent, surgically-created esotropia of 15 prism diopters and required prisms to fuse. Second, his head turn was not fully abolished. It was therefore decided to selectively weaken his left turners, that is, his right medial rectus muscle and his left lateral rectus muscle, doing proportionately more on the right medial rectus than on the left lateral rectus to overcome the esotropia and to shift his broadened null zone closer to the primary position. On 12/1/75, the right medial rectus muscle was recessed a further 2 mm. (a total recession of 7 mm.) and the left lateral rectus muscle was recessed 3 mm. (a total recession of 10 mm.).

Postoperatively, the esotropia was eliminated as a result of the second operation and the effect on the nystagmus intensity (Fig. 3) was to shift his null to the primary position.

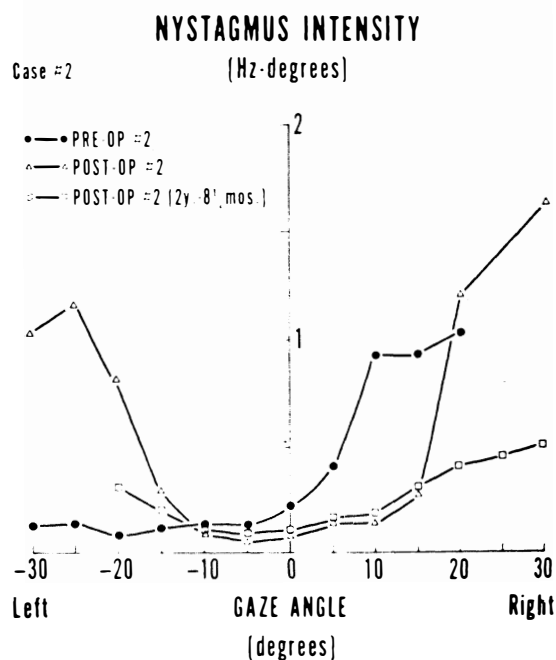


Fig. 3. Pre- and postoperative (procedure No. 2) plots of the nystagmus intensity vs. the gaze angle for Case 2.

On the last follow-up visit in July, 1978, the patient's binocular visual acuity in the primary position was 20/20, his monocular visual acuity was 20/25 in the right eye and 20/30 in the left eye. He had a slight (less than 5°) head turn to the right. His near binocular visual acuity was 20/20. On motility examination, the patient had, on cover test, an intermittent esotropia of 8 prism diopters at distance and 12 at near. His near point of convergence was remote, the right eye deviating. He had a large-amplitude, right-beating nystagmus on gaze to the right; on left gaze, he had limited ability to turn either eye to the left beyond 20° to 25° . Postoperative recordings continued to show a broad null zone centered on the primary position and diminished nystagmus intensity (Fig. 3).

This patient emphasized several management problems. The first of these was the creation of a tropia in a patient who was previously fusing. Although not emphasized in the literature on the surgery of congenital nystagmus, this occurs apparently about 20% of the time²¹. In this patient, it was accompanied by rapid partial recurrence of his head turn due to inadequacy of the surgery. Because of certain family problems, repeat surgery was postponed until some eighteen months after the first surgery. During the interim, the patient did reasonably well with a base-out prism over his right eye, which simultaneously helped him fuse and diminished his head turn to the right. The second surgical procedure has left him with an intermittent esotropia on cover test (the patient never experiences diplopia at the present time, however), but also has weakened his near point of convergence and constricted his gaze to the left. These are the trade-offs which one must make for effective surgery in congenital nystagmus. At the present time, we would avoid the so-called "5-6-7-8" operation²², as we think that it does not achieve the end of adequately shifting the null zone to the primary position.

Case Report 3. A 22 year-old white male born prematurely developed severe cicatricial retrolental fibroplasia which left him with essentially light perception in his right eye and reduced vision in his left eye. Nystagmus and a right esotropia appeared early in infancy. He had surgery on the right eye for the esotropia at age eight. The patient was aware that he had to turn his head to the left to see clearly with his left seeing eye.

Examination revealed a vision of light perception with projection in the right eye. In the left eye, the vision was 20/100 in the primary position and 20/60 in right gaze and less than 20/100 in gaze to the left. At near, his vision was 20/30 in the left eye in gaze to the right. Twenty prism diopters of right esotropia was present by prism corneal light reflex measurement. A poor direct light reflex and an afferent pupillary defect were

present on the right and a retrolental membrane with a peripheral traction detachment in the right eye. In the left eye, the anterior segment was normal, there was marked straightening of the retinal vessels, ectopia of the macula temporally, pigment changes and traction in the periphery consistent with a diagnosis of cicatricial retrolental fibroplasia.

Nystagmus recordings (Fig 4) demonstrated

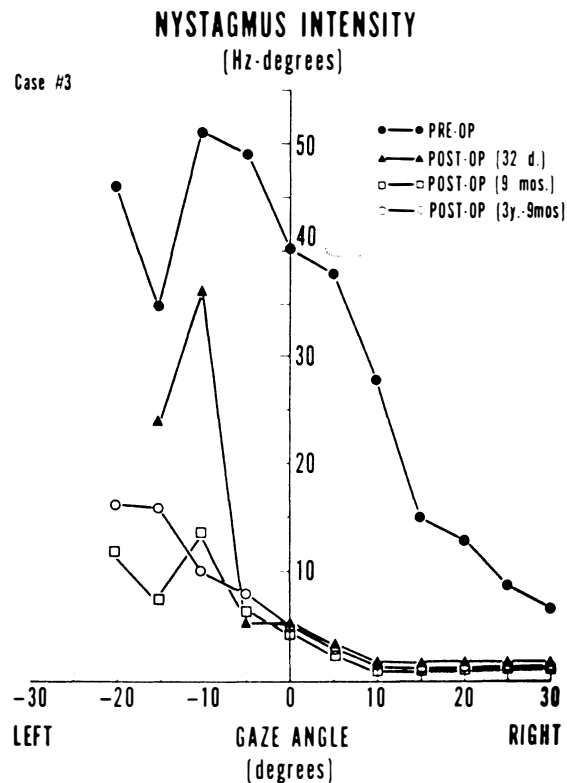


Fig. 4. Pre- and postoperative plots of the nystagmus intensity vs. gaze angle for Case 3, indicating the time course of improvement.

jerk nystagmus which decreased in intensity toward a null which was beyond 30° to the right.

The patient presented surgical indications similar to the Case Report 1 patient, but differed in that the null position was extremely displaced (beyond 30° to the right). This type of patient is a candidate for an immobilizing type of operation as described by Crone¹¹, i.e. a monocular patient with a null position in far eccentric gaze, the operation would of necessity overcorrect his esotropia. In addition, one could anticipate that the heterotopia of the macula would add to the real divergence that might occur postoperatively, a pseudodivergence due to his large positive angle kappa in the left eye. These problems were presented to the patient and because of the severity

of the cosmetic defect and his real desire for improvement of this, he decided to go ahead with surgery. A recession of the left medial rectus of 8 mm. combined with a recession of the conjunctival sac and resection of the left lateral rectus muscle of 7 mm. was performed on 12/9/74.

Postoperatively, much to our surprise, only a pseudodivergence due to the heterotopia of the macula occurred. The nystagmus intensity was markedly decreased (Fig. 4) and the null zone shifted to the left where it occupied a region between 10° and 30° to the right of the primary position. On last examination, the patient's nystagmus intensity and null zone have remained the same three years and nine months following surgery. The patient employs a head turn 12° to the left to see clearly. For him, this was enough cosmetic improvement and he does not wish to wear the required 20-25 diopter prism to compensate for it or undergo further surgery.

From the standpoint of the clinician, the problem presented by this patient was the enormous surgical requirement necessary to shift the left eye 30° or more to the left. In fact, inadequate surgery was performed and the patient was left with a 12° to 15° head turn. At the present time, our management of such a patient would be to do an adjustable medial rectus recession of 15 mm. or more combined with a recession of the conjunctival sac and a maximum later rectus resection. There is no question in our minds that such surgery is required in circumstances where the null zone is displaced more than 30° to the right or left of the primary position.

Case Report 4. The patient was a 28 year-old woman with congenital nystagmus and a right esotropia, poor vision in the right eye and a cosmetically disfiguring head turn to the left to obtain her best vision. The patient had lost several jobs because of the appearance of head turn and desired cosmetic correction of this.

On examination, the visual acuity in her right eye was 20/100 at distance and near. In her left eye, the visual acuity with a head turn 35° to the left was 20/50. In the primary position, the patient's vision was 20/100-. On clinical examination of her nystagmus, it seemed that her quiet phase in the left eye was about 15° to the right of the midline. In this position, it appeared that to further dampen her nystagmus, the patient would spontaneously develop an adduction movement of the right amblyopic eye of 15° to 20°.

On quantitative recordings (Fig. 5), she was noted to have a null zone in her left eye at about 10° to 15° of right gaze. Convergence of the right eye was also noted to dampen her nystagmus.

On 3/10/75, the patient had a left medial rectus recession of 5 mm., a left lateral rectus resection of 5 mm. and a right medial rectus recession of 6 mm. In the immediate postoperative period, al-

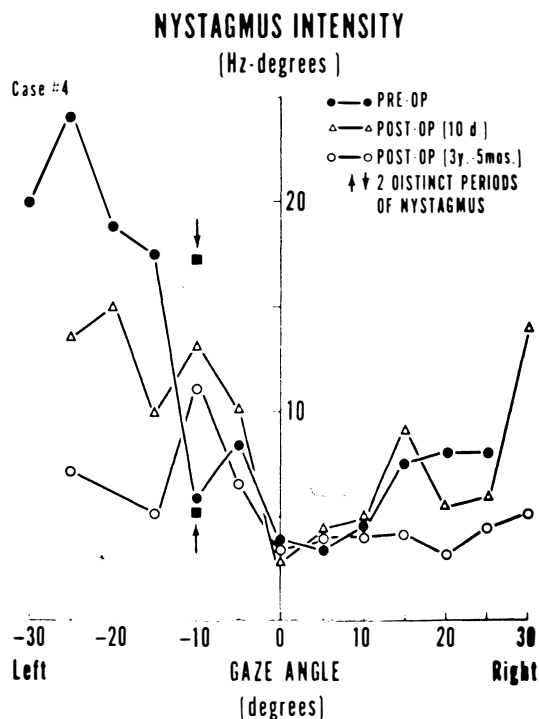


Fig. 5. Pre- and postoperative plots of the nystagmus intensity vs. the gaze angle for Case 4.

though her head turn to the left was diminished, it recurred as well as the tendency for her right eye to go esotropic when the left eye assumed a position of about 15° to the right of the primary position. When last seen on 8/10/78, the patient's visual acuity in her right eye was 20/100- and in her left eye 20/20-. She had a head turn of 15° to the left, best seen on determining her visual acuity at both distance and rear. The amblyopic right eye continued to go esotropic in varying amounts depending upon the effort the patient was making to obtain the best visual acuity from her left eye.

In re-evaluating this patient's eye movement recordings, it was obvious that she was not a simple, straightforward case of congenital nystagmus on the basis of her waveforms (Fig. 6). She actually was a case of combined congenital and latent nystagmus. These patients, in addition to their CN waveforms, have a completely different wave-form diagnostic of latent nystagmus²³. They represent an entirely separate entity in terms of treatment and are not amenable to the standard Anderson-Kestenbaum operation or its modifications. During the early years of our experience with quantitative recordings, we were unaware of this difference and did an operation which was of doubtful benefit to the patient.

Case Report 5. The patient was a 24 year-old black male first seen in the Eye Clinic on 2/16/76

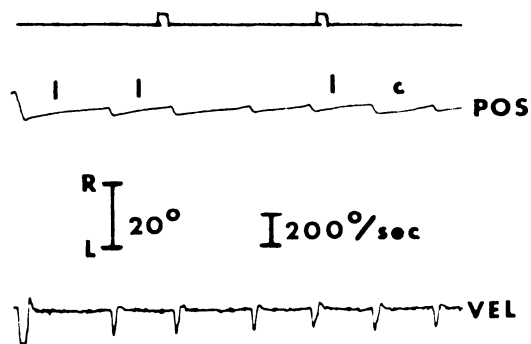


Fig. 6. A sample of the waveforms of Case 4 indicating the decreasing velocity slow phases diagnostic of latent nystagmus.

for complaints of headache and wiggling eyes. The patient's history of nystagmus goes back to early infancy and he claimed that he never had good vision in either eye. Neurological and medical evaluation revealed only mild diastolic hypertension.

On examination, the patient's visual acuity binocularly was 20/20; monocularly, it was 20/50-2 in the right eye and 20/25-1 in the left eye. His near visual acuity was 20/25. He had a marked head turn to the left of 25° to 30° . His nystagmus was jerk left in all fields of gaze including far to the right. His nystagmus dampened on convergence. The visual fields and fundus examination were normal.

On quantitative nystagmus recordings (Fig. 7), the patient had a null between 15° and 20° of right gaze.

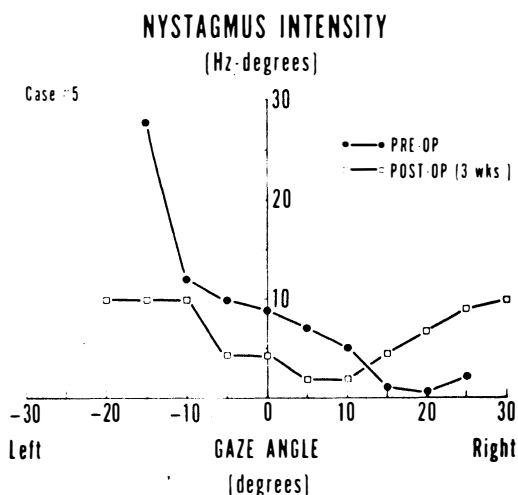


Fig. 7. Pre- and postoperative plots of the nystagmus intensity vs. the gaze angle for Case 5. The postoperative curve is based on limited data as explained in the text.

On 2/21/77, the patient had an Anderson-Kestenbaum operation performed, recessing his right lateral rectus muscle 11 mm., his left medial rectus muscle 6 mm. resecting his right medial rectus muscle 6 mm. and his left lateral rectus muscle 11 mm.

Postoperatively, his visual acuity was 20/20 monocularly and binocularly in the primary position at distance and near. He was orthophoric and had fusion for the Bagolini lenses at both distance and near. The patient was noted, however, to have large saccadic eye movements, horizontal in direction, random in occurrence and present primarily when he was being observed. These subsided spontaneously when the patient believed he was not being observed and when he put on dark sunglasses in order to read the visual acuity chart. The presence of these large saccades postoperatively (Fig. 8) prevented accurate quantitative re-evaluation of his nystagmus and the effect of the surgery on it. The curve shown in Fig. 7 is our best estimate based on the available data.

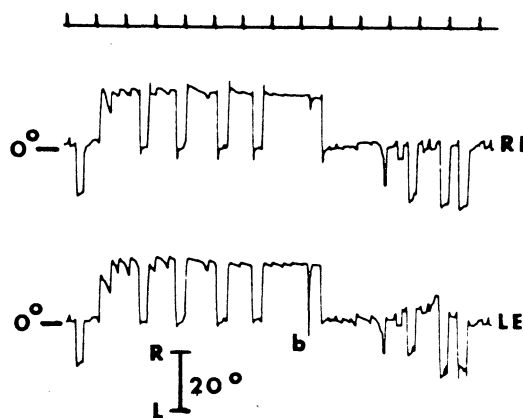


Fig. 8. Large saccades present postoperatively when the patient thought he was being observed.

The patient's behaviour postoperatively took us by surprise. However, review of his Jackson Memorial Hospital chart revealed numerous visits to the Emergency Room for trivial complaints. A diagnosis of hysteria and/or hypochondriasis was made on a number of occasions. Clearly, this patient physiologically benefitted from his Kestenbaum operation in the sense that he could now read the visual acuity chart with his head straight. This, however, for psychological reasons became intolerable for him. He substituted for his congenital nystagmus, large-amplitude, irregular saccades. These corresponded to no known pathological disorder of ocular motility. The problem presented by this patient in management was our failure to anticipate the effect of the surgery and the change that it wrought on the patient's psyche.

Postoperatively, the patient complained bitterly

of inability to control his eyes and had absolutely no insight into the fact that he was causing the very eye movements he could not control. Though contacted a number of times to return for follow-up evaluations and visits, the patient has steadfastly refused to keep these appointments.

Case Report 6. The patient is a 13 year-old white male who was noted shortly after birth to have a tendency to turn his head to the left. Nystagmus was noted shortly after that. There was no family history of serious eye problems.

On examination, his visual acuity binocularly was 20/40 with his head turned 20° to the left. His monocular visual acuity was 20/50-1 in the right eye and 20/200 in the left eye. At near, his visual acuity was 20/40 binocularly; monocularly, he was 20/50 in the left eye and 20/50 in the right eye. His nystagmus showed a large null zone from close to the primary position to 20° to 30° to the right of the primary position. His nystagmus elsewhere was jerk right in gaze to the right and jerk left in gaze to the left. It was left-beating in up gaze and down gaze. His nystagmus was dampened on convergence. He was orthophoric. The remainder of his ocular examination was normal.

On quantitative recordings (Fig. 9), the patient had a null zone of 20° to 25° to the right of the primary position. He also dampened his nystagmus significantly with convergence. On 12/19/77, a recession of the right lateral rectus muscle 10 mm.,

Postoperatively (3/30/78), his binocular visual acuity was 20/20. His monocular visual acuity was 20/30- in the right eye and 20/20 in the left eye. His head turn appeared to be about 10° to the left. Postoperative recordings on that same day (3/30/78) showed diminished nystagmus intensity and a broad null zone encompassing the primary position (Fig. 9). The absolute null at 10° right gaze corresponded to his 10° left head turn and, more importantly, to an area in which longer periods of motionless foveation occurred per cycle and allowed extremely good vision.

This patient also presented the classical indications for the Anderson-Kestenbaum operation. This operation diminished his nystagmus intensity, broadened his null zone and shifted it to the primary position. In addition, his binocular visual acuity improved as a result of these changes in the characteristics of his nystagmus. He still has a slight head turn to the left but, according to the patient and his family, this is much improved over what it was and acceptable to him and them.

DISCUSSION

We have presented the results of our experience with six patients in whom quantitative recordings of their nystagmus could be performed both pre- and postoperatively. The surgical approach to congenital nystagmus produces the following changes: the nystagmus intensity is diminished over practically all gaze angles. The null zone is generally broadened and shifted toward the primary position. A spontaneous improvement in the visual acuity may occur as a result of these changes.

Clinically, our six patients presented varying problems in management. The first patient represented the application of the Anderson-Kestenbaum technique to the monocular patient with strabismus and amblyopia. Postoperatively, her head turn recurred but this was easily handled by a prescription of a small prism. The second patient represents two complications of congenital nystagmus surgery, namely, the creation of a tropia where none existed before and the rapid recurrence of the head turn. Both of these proved amenable to reoperation. Our current practice is to reoperate these patients within three months if the head turn recurs or a tropia is created as a result of the surgery. The third patient presented indications similar to the first but, because of the location of his null zone far in right gaze, he required an immobilizing operation to shift his null zone to the primary position. Inadequate surgery was performed and the head turn, although cosmetically acceptable to the patient, is still in the region of 12°. The fourth patient represents an example of latent nystagmus in conjunction with true congenital nystagmus with a head turn. This type of patient is not usually amenable to congenital nystagmus

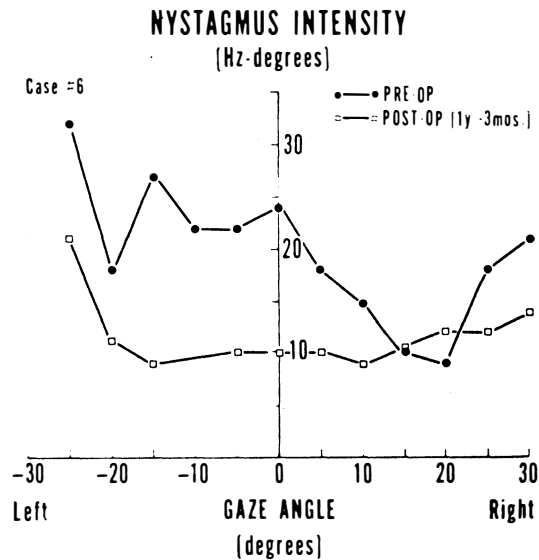


Fig. 9. Pre- and postoperative plots of the nystagmus intensity vs. the gaze angle for Case 6.

a resection of the right medial rectus muscle 6 mm., a recession of the left medial rectus muscle 6 mm. and a resection of the left lateral rectus muscle 10 mm. was performed.

surgery. Clinically, these patients can appear very similar to ordinary congenital nystagmus patients; however, on recordings of their movements, they show a waveform characteristic of latent nystagmus (a decelerating exponential slow phase) (Fig. 6) in addition to those of true CN. As one would expect, she did not benefit significantly from the congenital nystagmus surgery. The fifth patient represents a clear-cut illustration of the truism that the physician is responsible in his treatment for the whole patient, not just his eyes. We clearly missed the many dependency-created psychological problems that this patient manifested in his other medical history. Simple review of his medical record prior to the surgery would have revealed this to us. Although the nystagmus surgery itself was successful, it was quickly replaced by the patient with an even more crippling psychologically-induced type of bizarre eye movement which, according to the physicians who have seen the patient, persists to this day. The sixth patient represents the happy results which can occur in congenital nystagmus surgery, both clinically and in the eye movement laboratory.

Although the field of congenital nystagmus surgery has been expanding for the last twenty-five years since its introduction by Anderson and Kestenbaum, there is still a great deal to be learned in applying their principles to the patient with congenital nystagmus. The use of quantitative pre- and postoperative recordings has proven valuable in our hands in fully documenting the nature of the nystagmus, in confirming the presence of a null zone, measuring its distance from the primary position, in planning the surgical approach and, finally, in quantitatively evaluating the results of surgery. They have documented clearly those cases where the surgery was slightly inadequate and resulted in an improved but not abolished head turn. The stability of the measured postoperative null positions should dispel the current popular notion that the head turn (i.e. the off-primary position null angle) "recurs" at some variable time postoperatively. If the surgery was insufficient the null position will be seen at its new, less severe position immediately postoperatively and remain fixed.

BIBLIOGRAPHY

1. Anderson, JR: Causes and treatment of congenital eccentric nystagmus. *Brit J Ophthalmol* 37:267-281, 1953.
2. Kestenbaum, A: Nouvelle operation de nystagmus. *Bull Soc Ophtal Fr* 6:599-602, 1953.
3. Goto, N: A study of optic nystagmus by the electro-oculogram. *Acta Soc Ophthal Jap* 58: 851-865, 1954.
4. Pierce, D: Operations on vertical muscles in cases of nystagmus. *Brit J Ophthalmol* 43: 230-233, 1959.
5. Sternberg-Raab, A: Anderson-Kestenbaum operation for asymmetric gaze nystagmus. *Brit J Ophthalmol* 47:339-345, 1963.
6. Cooper, EL, Sandall, GS: Surgical treatment of congenital nystagmus. *Arch Ophthalmol* 81:473-480, 1969.
7. Sandall, GS, Cooper, EL: Surgical therapy of congenital jerk-type nystagmus. Its cause and effect on heterotropia. *Doc Ophthalmol* 34: 361-364, 1973.
8. Schlossman, A: Nystagmus with strabismus: Surgical management. *Trans Am Acad Ophthalmol Otolaryngol* 76:1479-1486, 1972.
9. Schlossman, A: The effect of nystagmus on comitant strabismus: Surgical implications. *Ann Inst Barraquer* 12:341-353, 1974-5.
10. Taylor, JN: Surgery for horizontal nystagmus: Anderson-Kestenbaum operation. *Aust J Ophthalmol* 1:114-116, 1973.
11. Crone, RA: The operative therapy of nystagmus. *Ophthalmologica* 163:15-20, 1971.
12. Friede, R: Zur operativen Behandlung des Nystagmus gravis nebst Bemerkungen zur Extraction der kongenitalen Katarakt. *Klin Monatsbl Augenheilkd* 128:451-455, 1956.
13. Kommerell, G: Nystagmusoperationen zur Korrektur verschied ener Kopfwangshaltungen. *Klin Monatsbl Augenheilkd* 164:172-191, 1974.
14. Parks, MM: Congenital nystagmus surgery. *Am Orthopt Jr* 23: 35-40, 1973.
15. Calhoun, JH, Harley, RD: Surgery for abnormal head position in congenital nystagmus. *Trans Am Ophthalmol Soc* 71:70-87, 1973.
16. Hugonnier, R: Notre Position Actuelle sur les Operations Dirigees Contre le Nystagmus. *Ann Inst Barraquer* 12:327-340, 1974-5.
17. Fells, P, Dullely, B: Surgical management of compensatory head posture. *Trans Ophthalmol Soc UK* 96:90-95, 1976.
18. Dell'Osso, LF, Flynn, JT: Congenital nystagmus surgery: A quantitative evaluation of the effects. *Arch Ophthalmol*. In press.
19. Dell'Osso, LF, Flynn, JT, Daroff, RB: Hereditary congenital nystagmus: An intrafamilial study. *Arch Ophthalmol* 92:366-374, 1974.
20. Dell'Osso, LF: Fixation characteristics in hereditary congenital nystagmus. *Am J Optom* 50:85-90, 1973.

21. Rosenbaum, AL, Metz, HS, Carlson, M, Jampolsky, AJ: Adjustable rectus muscle recession surgery. Arch Ophthalmol 95:817-820, 1977.
22. Calhoun, JH: Personal communication.
23. Daroff, RB, Troost, BT, Dell'Osso, LF: Nystagmus and Related Ocular Oscillations, in "Neuro-Ophthalmology," J.S. Glaser (ed.), Harper & Row, Hagerstown, Md., chap 11, 1978.

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