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Surgery of congenital nystagmus

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Summary
Congenital nystagmus surgery is discussed from the standpoint of measuring quantitatively the effects of surgery on the waveform, amplitude, and frequency of the nystagmus oscillation.

Congenital nystagmus (CN) surgery, advocated independently and simultaneously by Anderson and Kestenbaum in 1953 (Anderson, 1953; Kestenbaum, 1953) for the relief of cosmetically disfiguring head positions associated with the nystagmus, has been shown to have wider applications (Pierce, 1959; Cooper and Sandall, 1969; Sandall and Cooper, 1973; Schlossman, 1972) than that envisioned by these pioneers and to produce effects not originally contemplated by them (Taylor, (1973; Crone, 1971; Flynn and Dell'Osso, 1979; Dell'Osso and Flynn, 1979).

In this communication, we hope to bring to our readers attention the clinical applications of certain technical advances to the observation of the behaviour of the eye with CN which have broadened and deepened our understanding of what happens in that eye as it rhythmically oscillates under the nystagmus impulse. These observations underlie the rationale of the surgical approach to CN. Their limitations point to fruitful areas for further clinical research aimed at penetrating deeper into this fascinating clinical problem.

I. Fixation behaviour

A first step for the ophthalmic surgeon planning a surgical approach to the patient with CN is the observation of the *fixation behaviour* of the fovea by direct visualization of the fundus. The feasibility of this technique was cinematographically recorded (Dell-Osso, 1973) employing fundus contact lens, slit lamp, beam splitter, and movie camera. An important conclusion for nystagmus surgery from analysis of these films was the fact that the fovea, regardless of the waveform of the nystagmus, was always at the velocity minimum of the wave. The brain, in effect, employed a strategy to ensure that the object of regard would be localized on the fovea when the speed and direction of each individual nystagmus wave was at its minimum. The essence of surgery for CN is to reinforce and support this effective strategy.

For the ophthalmic surgeon contemplating surgical intervention today, such recording apparatus is neither an appropriate nor a necessary part of his examination. His observation of the fixation behaviour of the eye is, however, essential. For this he needs to employ an ordinary ophthalmoscope with a fixation reticle and to observe fixation in at least the field of the 'null zone' (Kestenbaum, 1961), in the primary position, and in the field where the nystagmus appears greatest. Uniocular and binocular viewing is helpful. This simple physiological observation, demonstrating a definite dampening of the oscillation in its preferred field of gaze, should be sought in each patient considered for surgery.

II. Eye movement recordings

The advances in techniques of recording eye movements accurately in the years since 1953 have been impressive (Alpern, 1969). Unfortunately, most of the applications of these advances have been limited to the physiological laboratory rather than the clinic. The authors have made a systematic attempt to study CN patients by infrared reflection techniques both pre- and postoperatively. To present the results of such recordings, the concept of nystagmus intensity (Dell'Osso, Flynn, and Daroff, 1974) has been introduced. This derived quantity is simply the product of the nystagmus frequency in cycles per second multiplied by the peak-to-peak amplitude of the waves in degrees. This product is plotted for each gaze angle across the horizontal meridian (or any other meridian chosen) as the subject views a light-emitting diode on the arc of a

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modified perimeter (Fig. 1). It illustrates at a glance the null zone location, breadth, and depth as perhaps no other method can. The shift of this null zone to the primary position is the goal of surgery. The advantages of this method of quantitative measurement are obvious, particularly as it permits easy comparison between pre- and postoperative nystagmus intensity recorded under the same conditions. The technique has permitted the demonstration of the stability of the results of CN surgery over long periods of time in the small group of subjects studied (Flynn and Dell'Osso, 1980). Its chief disadvantage is the paucity of the oculomotor laboratories on either side of the Atlantic that have the capability to perform such recordings.

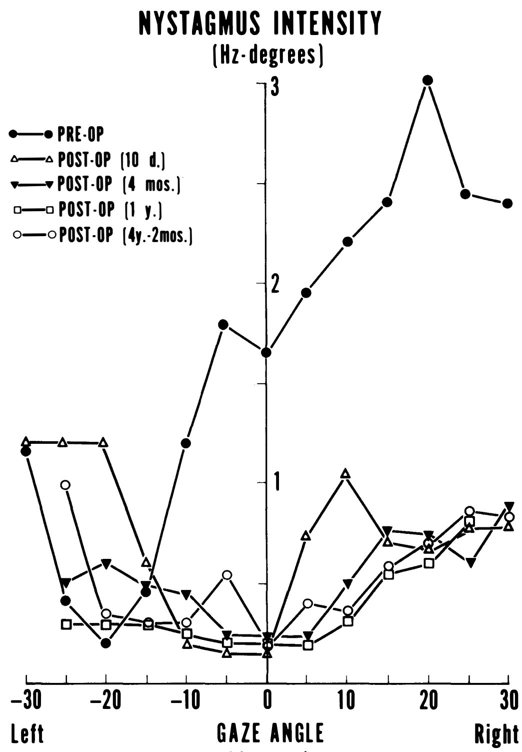

FIG. 1 *Case 1. Nystagmus intensity of Hertz (cycle/second) degrees plotted as a function of gaze angle. This measure is made both pre- and postoperatively in a patient who has had surgery for congenital nystagmus*

One of the immediate results of this form of study of CN intensity plotted in this fashion has been the recognition that CN surgery, when effective, not only shifts that null zone towards the primary position but also may broaden and deepen it (Fig. 2). This latter effect, reduction of nystagmus intensity within the null zone, is probably responsible for the spontaneous improvement in visual acuity noted in some patients after CN surgery. The method further demonstrated

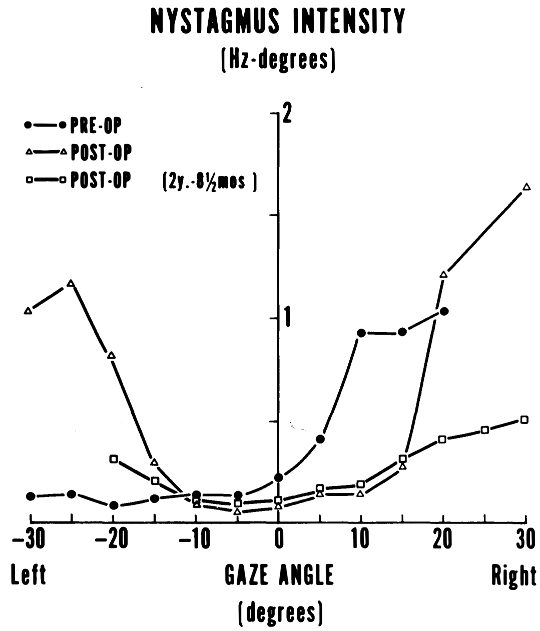

FIG. 2 *Case 2. Nystagmus intensity plot to demonstrate the shift of the null zone to the primary position with reduction in the nystagmus intensity, particularly notable in the region of the primary position.*

that nystagmus intensity diminished across the range of gaze angles recorded away from the shifted null zone and appeared to remain reduced over significant follow up periods.

III. Effect of surgery on waveform-frequency-amplitude

The type of nystagmus intensity plot described above is useful in quickly grasping the major effects of CN surgery thus far observed. It is clear, however, that nystagmus intensity is a derived quantity reflecting the product of two putative independent variables, frequency and amplitude. As such, information about either is lost at any gaze angle in plotting their product. Yet it seems apparent that this information and that of the third variable, waveform, may be critical in further assessing the useful long-term effects of CN surgery. As we indicated at the outset, it appears that the central nervous system, faced with the problem of attaining the best possible vision with a constantly oscillating eye, is employing a strategy aimed at maximizing the time spent by the object of regard on the fovea. Surgery performed for CN should, as far as possible, conform to that strategy.

With this in mind, we recently reviewed the recordings of two patients. The questions we asked were: 'Are waveform, frequency, and amplitude all affected by CN surgery?' 'If not, which are and to what extent?' The preliminary answers are contained in the Table. Waveform, as not unexpected, is resistant to the effects of CN surgery in all cases studied thus far.

Table *Nystagmus parameters*

Case No.	*Preoperative at null zone*			*Postoperative at primary position*			*Ratio preop/postop*	
	Amplitude (°p–p)	*Frequency (cps)*	*Wave-form*	*Amplitude (°p–p)*	*Frequency (cps)*	*Wave-form*	*Amplitude*	*Frequency*
1	1	2	J*	0.5	2	J	0.5	1
2	0.5	2	Jef†	0.5	1	Jef	1	0.5

*J = Jerk waveform † Jef = Jerk waveform with extended foveation

Amplitude and frequency are not. Either or both can be affected by surgical intervention.

But there appears no clear-cut correlation between the type of surgery performed and the different effects achieved

Discussion

In this report, we have attempted to focus attention on surgical indications and technique (Crone, 1971; Friede, 1956; Kommerell, 1974; Parks, 1973; Calhoun and Harley, 1973; Fells and Dulley 1976; Hugonnier, 1974–5), but rather to probe beneath the surface of the obvious beneficial effects of Anderson-Kestenbaum surgery in an attempt to understand the mechanism of its benefits. Employing 'state of the art' recording techniques has demonstrated compellingly that the head posture assumed by the patient with CN maximizes the benefit of his foveation strategy by damping as far as possible the nystagmus frequency and amplitude. Nystagmus surgery has been shown to have a general effect on nystagmus intensity across a range of gaze angles as well as shifting the null zone to the primary position. Within the null zone itself, further reduction of nystagmus intensity permits, in some cases, an improvement in the visual acuity. Finally, a preliminary analysis of the effects of CN surgery on the three parameters which specify nystagmus oscillation—waveform, amplitude, and frequency—reveals that the first is insensitive to surgery but the two latter are affected in unpredictable ways.

Our purpose should be clear to our readers. More quantitative and analytical work is clearly needed in the field of CN surgery. The use of different visual targets, different viewing conditions, the possibility of combining pharmacological effects (Halmagyi, Rudge, and Gresty, 1980) with surgical therapy in selected cases, and the study of the effects of newer operative techniques such as the Faden operation (Mühlendych, 1975) are but a few of the channels open to the surgeon with an interest in studying this problem in his patients.

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