(Accepted 24 August 1986)

Evaluation of smooth pursuit in the presence of congenital nystagmus

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ABSTRACT. Studies of the ability of patients with congenital nystagmus (CN) to pursue a moving target have yielded conflicting conclusions. This paper reviews pursuit records dating from the first quantitative eye movement study of smooth pursuit and CN (with its hypothesis of superposition of normal pursuit onto the CN) to more recent records obtained when such patients pursue a moving target. Correct interpretation of the resulting ocular motility data is dependent upon the understanding of CN waveform and direction changes, null shifts and target foveation that has emerged in the 20 years since that first study. The data support the conclusion that the smooth pursuit system in CN patients is intact, using both retinal slip velocity and position information to match eye velocity and position to those of the target. An updated model of the author's original hypothesis is included to illustrate how this is possible. Thus, the mechanisms used are identical to those employed by normals and differences in the resulting eye movements are due to the presence of the ever-changing CN waveform superimposed upon normal efferent pursuit commands.

Key words: smooth pursuit; eye movements; congenital nystagmus

INTRODUCTION

The normal smooth pursuit system causes the eyes to move with a velocity equal to that of the target being pursued

Most, if not all, researchers in the field of ocular motility would accept the above statement as a reasonable definition of normal smooth pursuit. Yet, it almost always represents an incorrect oversimplification. Most currently accepted models of the ocular motor system include a summation of neural control signals at the ocular motor nuclei (OMN). The signals converging on the OMN include: position and velocity from the saccadic, pursuit and vestibulo-ocular systems; vergence; and other, spontaneous oscillations and drifts. The oscillations may arrive via the former pathways or some additional, as yet undefined pathways. Thus, in general, although the *output* of the normal pursuit system is a velocity signal equal to target velocity, actual *eye velocity* equals that pursuit velocity summed with any other velocity signals

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from the saccadic, vestibulo-ocular or vergence systems plus any spontaneous velocities, such as the slow phase velocity of nystagmus. Only under the unique condition when *all* other velocity signals are identically zero, will the eye velocity match the target velocity. It is this very simple, but important, point that seems to have been overlooked by many who have reported on smooth pursuit in the presence of spontaneous nystagmus.

The first quantitative studies of smooth pursuit in a subject who had congenital nystagmus (CN) were carried out in an electrical engineering laboratory at the University of Wyoming in 1965 (Dell'Osso, January 1968). In retrospect it was serendipitous that the particular CN subject studied had a pendular form of CN rather than a jerk nystagmus. The actual waveform was pendular with foveating saccades but at the time of the study the various waveforms of CN were unknown, as was the concept of target foveation during a particular portion of a CN waveform. It was fortuitous that a pendular waveform was studied first since it enabled the experimenter to correctly interpret the data and not be misled by the obligatory saccades that are a part of all jerk waveforms. No such saccades were present in the response and it was observed that the eye movements produced by a person with pendular CN consisted of that same pendular oscillation riding on a change in eye position that followed exactly the target motion. This was hypothesized to be the result of a normally operating smooth pursuit system adding its output to the internally generated CN oscillation. The additive response was recorded for targets moving with constant velocity (ramps) and for those moving in a sinusoidal (pendular) manner. This hypothesis was embodied in a model of the ocular motor system that contained both a saccadic and smooth pursuit system and that duplicated the pulse, step, ramp and step-ramp (Rashbass) responses of both normals and the patient with pendular CN (Dell'Osso, January 1968, 1967, 1970). Included in the model was: an efference copy pathway to allow the model to ignore movements caused by the oscillation (*i.e.*, the model, like the patient, had no 'oscillopsia') and act on true changes in target position and velocity; the variation with gaze angle of the CN amplitude; and, the nulling of the CN at a gaze angle equal to that of the patient.

In a study prompted by the above work, it was found that the CN patient had normal saccades and better than normal smooth pursuit (Dell' Osso *et al.*, 1972). The normal saccades implied that both the ocular motor control signals and extra-ocular muscular system were normal. The pursuit was characterized by an ability to track more rapidly moving targets than normals could and to track in phase with the target at higher frequencies than normals. The data also showed, for the first time, that the superimposed CN was influenced by the direction of the smooth pursuit as well as by eye position; this latter observation was to prove extremely important in the analysis of the smooth pursuit of patients with jerk CN waveforms and is responsible for clarifying the mistaken impression that CN patients respond to optokinetic stimuli with 'reversed' optokinetic nystagmus (OKN) (Halmagyi *et al.*, 1980). In order to fully understand the mechanisms involved in the pursuit of a moving target by an oscillating ocular motor system, it is imperative to first clarify those involved in fixation (foveation) of a stationary target; this is in keeping with the idea, first proposed by Yarbus, that fixation is equivalent to pursuit at zero velocity (Yarbus, 1967). Therefore, the results of studies of CN fixation will be discussed first with particular attention paid to their relevance to smooth pursuit.

METHODS

The methods used in studying the eye movements of CN patients vary from the home-made, direct current, white light reflection technique used in that first quantitative study in 1965 to infrared reflection, scleral search coils, and even electrooculography (EOG). They are all well described in the sources referenced herein and will not be further discussed except to remind the reader that EOG is the method most subject to error (due to drifts and noise) and least sensitive. References made in this paper to 'quantitative' studies of eye movements refer to the above photo-electric or electromagnetic methods but do not include EOG.

INITIAL STUDIES OF CN FIXATION

The above studies and modelling of smooth pursuit and CN were done before anything was known about CN waveforms and their relation to target foveation. In fact, to minimize position error the oscillatory eye movement tracings were positioned on the chart paper so they straddled the grid lines representing target position. Although this coincided with the existing clinical descriptions of CN it was clear to me even then that such descriptions were incorrect since they precluded the good visual acuity known to exist in many CN patients. Before the hypothesis represented in the model could be studied further, more had to be known about the relationship of the oscillating eye and the stationary target being fixated; this was essential for accurate calibration.

Using a laser target and retinal cinematography we established that, for both pendular and jerk waveforms of CN, the eyes oscillated in a manner that brought the fovea away from and back to the target (Dell'Osso, 1973). Thus, the target was imaged on the fovea at one peak of the oscillation when the eye motion was reversing direction and velocity was at, or near, zero. This discovery was the key to understanding how the visual acuity of CN patients could be normal despite the large oscillations. More recent studies have confirmed our initial observations. Using an infrared TV fundus camera, Tani (1981) found the same foveation periods in CN waveforms. Also, measurements of the stability of CN foveation periods have revealed standard deviations of only 25 to 65 minarc (well within the foveal area) for different patients (Bedell *et al.*, 1985). Thus, despite CN oscillations of several degrees peak-to-peak amplitude, the *fixation reflexes* of these patients are remarkably *good.* When we combined this new knowledge with accurate tracings produced using infrared reflection, we were able to identify 12 distinct waveforms and their relationship to target position (Dell'Osso & Daroff, 1975). Many of these waveforms contain slow phases distorted by the patient's fixation reflexes to produce long periods of foveation (up to several hundred milliseconds) during each cycle of the CN. It soon became easy, with a little practice, to identify both target foveation and spontaneous changes in the direction of the instability (bias reversals) (Dell'Osso, 1973) that occurred at any given gaze angle for a particular patient.

The picture of CN that emerged was that of an instability that manifested itself in either pendular or increasing-velocity runaway from the target in a direction that was idiosyncratic and depended on the position of the static null angle or the static neutral zone (SNZ) of the CN. The neutral zone is defined as the zone where the nystagmus reverses direction (Kestenbaum, 1948). The gaze angle at which the CN nulls or where the direction of a jerk nystagmus reverses (the same for almost all patients) determines the

CN direction at any other gaze angle. If a target is viewed at an angle to the left of the null the CN will be jerk left, to the right, the CN will be jerk right. Further away from the null, the CN amplitude increases due to more rapidly accelerating slow phases with little or no periods of extended foveation. The static null angle (*i.e.*, when fixating stationary targets) is fixed for a particular patient (Dell'Osso, 1984). For some patients it could be shifted by covering either eye; this is referred to as a latent component of CN and is *not* latent nystagmus (Kestenbaum, 1940, 1961; Daroff *et al.*, 1973; Daroff & Dell' Osso, 1974; Dell'Osso, 1982a, 1982b).

As we observed in the study described above (Dell'Osso *et al.*, 1972), the null position could also be shifted by smooth pursuit. Subsequent recordings of many other patients revealed that the null shift (or position of the 'dynamic null') was a function of both pursuit direction and velocity (Halmagyi *et al.*, 1980; Dell'Osso, 1982a, 1982b, 1984). The dynamic null shifted in the direction opposite to the pursuit direction and by an amount related to pursuit velocity. Thus, recordings of CN patients pursuing to the left, showed both the null and neutral zone appearing at a gaze angle to the right of their static null and *vice versa.* The null shift is the cause of the obligate direction reversal of CN over a range of gaze angles that depends on the amount of that shift. It is this complex interaction, of smooth pursuit and the dynamic equilibrium of the unstable ocular motor system causing CN, that is responsible for the confusion evident in several recent papers on the subject.

To summarize the salient points of CN fixation that affect analysis of CN pursuit: foveation of stationary targets takes place during brief periods of time every cycle; such 'snapshots of vision' are enough for perception of a clear image of the world as well as good (even normal) acuity; the direction of CN is dependent on gaze angle and both pursuit direction and velocity due to the dynamics of the null position.

HYPOTHESIS AND EXPECTATIONS

What is meant by the application of the word 'normal' to both fixation and pursuit of ocular motor systems when the resulting eye movements certainly do not appear 'normal'? A record of fixation of a stationary target by a normal subject would show a straight line of constant eye position (zero velocity). Thus, normal fixation implies: eye position = target position and eye velocity = 0 (physiological drifts, flicks and tremor excepted). Clearly this cannot be expected of a CN patient, whose eyes are in constant motion, except at those short foveation periods described above. Thus, CN fixation (exemplified by the sketch in Fig. 1a) is not 'normal' but careful analysis of eye movement records reveal that fovation periods line up to form a straight line, as indicated by the dashed line of Fig. 1a. Such accuracy indicates that, despite the ongoing oscillation, the patient's fixation reflexes are indeed normal. The eye position during the foveation periods of each beat is such that the target is imaged directly on the fovea (*i.e.*, there is little or no 'jitter' in the recording). For the CN patient, therefore, it is reasonable to define this as 'normal' fixation. Some CN patients with foveal abnormalities or other sensory defects do not exhibit such stable foveation periods (*i.e.*, the recording has jitter) and are not able to fixate normally independent of the CN. In the presence of instabilities such as CN, one must carefully define *normalcy of function*, given the basic abnormality present, and clearly understand what may reasonably be expected from

normal ocular motor subsystems under the circumstances in order to avoid the trap of simply declaring all responses to be abnormal and thereby shedding no light on the operation of these subsystems. This paper will show that the smooth pursuit system of CN patients functions normally and, using the same arguments, one can infer that both the vestibular and optokinetic systems do also.

If Fig. 1a represents normal CN fixation (all foveation periods line up on-target) resulting in stable perception of a target (no oscillopsia), what might we expect of a normal smooth pursuit system operating under the oscillatory conditions imposed by CN? We could postulate that the act of pursuit would cause all oscillatory movements to cease and the resulting eye movements should then appear as normal smooth pursuit; that seems highly unlikely although, as we will see below, under some circumstances certain patients exhibit just such behavior. A much more parsimonious expectation is that embodied in the model described above (Dell'Osso, January 1968). The normal smooth pursuit apparatus extracts and acts upon true retinal slip velocity since efference copy provides knowledge of the internally generated oscillatory movements (*i.e.*, the brain internally cancels the CN waveform to prevent oscillopsia). The resulting smooth pursuit eye movement consists of the superposition of the CN and normal smooth pursuit. Certainly that was what was observed for pendular CN and what resulted in the model hypothesis. The superposition of two such waveforms, each unchanging, results in the sketch shown in Fig. 1c. The waveform shown in 1c was constructed by the addition of the waveforms shown in 1a (normal fixation of a CN patient) and 1b (the output of a normal smooth pursuit system). At first glance, one might not consider Fig. 1c to

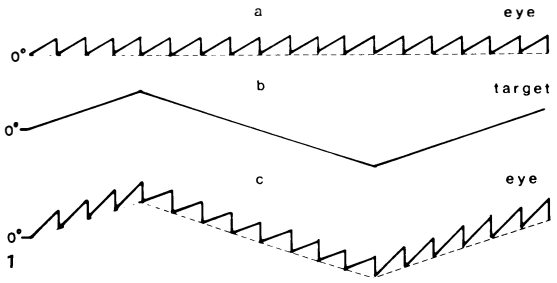

Fig. 1. Illustration of the relationship of any jerk left nystagmus to a target during (a) fixation of the stationary target and (c) smooth pursuit of the target moving as shown in (b). The dashed lines in (a) and (c) trace the patient's line of sight and represent fixation and pursuit respectively when the nystagmus is subtracted out. The eye movements in (c) are the result of summing the nystagmus of (a) with the target motion (or, the output of a normal smooth pursuit system) of (b).

represent the eye movements of someone with a normal smooth pursuit system. However, we have *generated* them by summing an ongoing, albeit unchanging, CN with normal smooth pursuit; they are, therefore, the eye movements resulting from normal smooth pursuit – by construction.

If, further, one remembers that a person with CN only foveates a target (whether it be stationary or moving) just after the fast phase, in the initial portion of the slow phase, one can consider the dashed lines in Fig. 1c as tracing this person's line of sight (that which he perceives) while fixating the moving target. Having done this, we can ignore the CN and concentrate on what the patient's line of perceptual sight is doing; we see that it is following the smooth pursuit target perfectly. This is analogous to the dashed line of Fig. 1a which represents the line of sight when the target was not moving. In both cases the ongoing CN is subtracted out and the results are stable fixation and good smooth pursuit, respectively. These dashed lines represent

the perception of the patient with CN when fixating either a stationary or a moving target (he does not have oscillopsia). Fig. 1a represents the ability to foveate a stationary target (normal fixation) and Fig. 1c represents the ability to foveally pursue a moving target (normal pursuit). The patient gets from 1a to 1c by superimposing the normal output from his smooth pursuit system onto his CN waveform. The message of Fig. 1 is that for nystagmus waveforms without flat foveation periods during fixation of stationary targets, one cannot use responses like Fig. 1c to infer poor smooth pursuit. Although the response may be generated by the saccadic system (which would infer poor pursuit) it could equally be due to superposition of normal smooth pursuit onto the ongoing nystagmus.

Fig. 2 is a more realistic sketch of a CN waveform when the patient is foveating a stationary target (2a) or pursuing a moving target (2c). Note that after the foveating fast phases are periods of motionless extended foveation in 2a and 'tracking foveation' in 2c; the periods of extended foveation in 2c have the same slope as the target motion shown in 2b (eye velocity = target velocity). Thus, the only time eye velocity reflects the *actual smooth pursuit output* of such a patient is during these otherwise motionless periods in the patient's waveform. Again, Fig. 2c is a sketch of a normal pursuit signal (2b) added to an *unchanging* CN (unfortunately, this latter condition is rarely observed). Because of the dynamic properties of the CN null discussed above, one should not expect such a simple picture as is shown in Fig. 2, although in certain cases this is exactly what the patient's eye movement records show. Responses similar to Fig. 2c are difficult to conceive of as being caused by any known ocular motor mechanism other than normal smooth pursuit; this is reinforced by the dashed lines of Fig. 2c that result from subtracting out the CN.

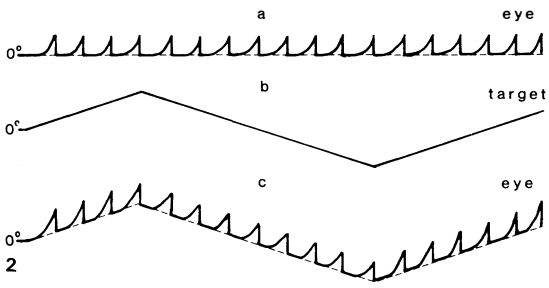

Fig. 2. Illustration of the relationship of a jerk left with extended foveation CN waveform to a target during (a) fixation of the stationary target and (c) smooth pursuit of the target moving as shown in (b). The dashed lines in (a) and (c) trace the patient's line of sight and represent fixation and pursuit respectively when the CN is subtracted out. The eye movements in (c) are the result of summing the CN of (a) with the target motion (or, the output of a normal smooth pursuit system) of (b).

Fig. 3 shows the effect of changing pursuit velocities on the resulting tracking record. As in Figs. 1 and 2, it is a simulation of a spontaneous nystagmus (3a) plus a normal smooth pursuit command (3b) yielding the pursuit eye movements (3c). There are some important observations that can be made from this figure that apply to the analysis of smooth pursuit in the presence of any spontaneous nystagmus, congenital or acquired. The first is that the velocity of the eye can *never* equal that of the target if there are no periods of zero velocity in the spontaneous nystagmus waveform; the hypothesis of superposition precludes this since the resultant eye velocity equals pursuit velocity plus slow phase velocity. Understanding this point is extremely important if one is to correctly analyze the smooth pursuit records of many different types of nystagmus patients. Put simply, one cannot presume, from the absence of smooth, continuous eye movements in a smooth pursuit record of a patient with spontaneous nystagmus,

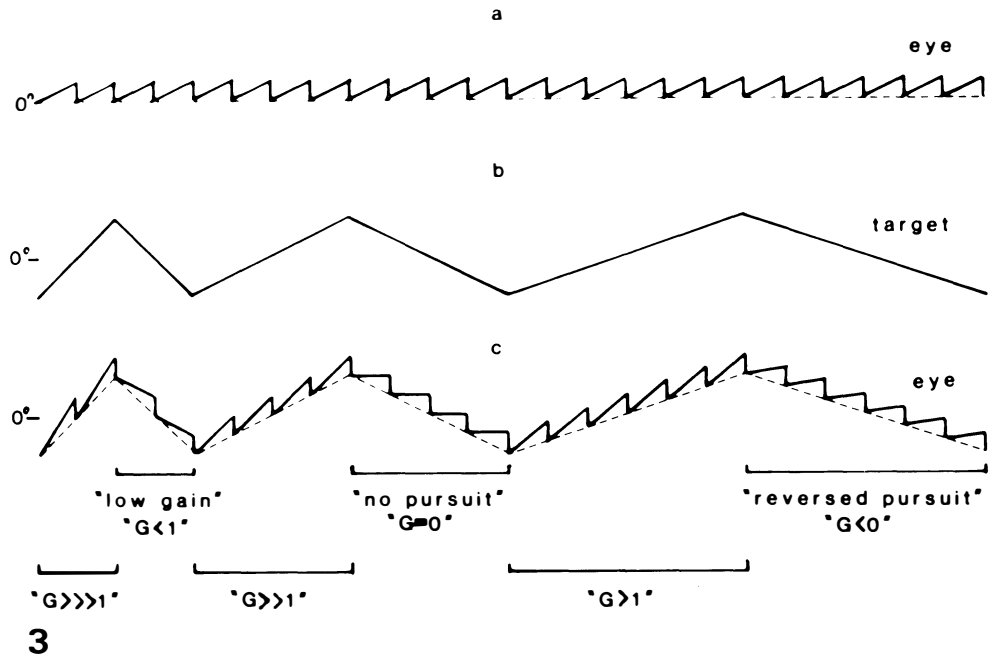

Fig. 3. The effects of changing pursuit velocity on the resulting eye movement record that contains nystagmus of any origin. Fixation of a stationary target is shown in (a) pursuit of a moving target in (c); the target motion (or, the output of a normal smooth pursuit system) is shown in (b). As in Figs. 1 and 2, the dashed lines in (a) and (c) trace the patient's line of sight and represent fixation and pursuit respectively when the nystagmus is subtracted out. The eye movements in (c) are the result of summing the nystagmus of (a) with the varying target motion (or, the normal smooth pursuit output) of (b). Pursuit gains 'G' and pursuit descriptions are placed in quotation marks to indicate the erroneous nature of these conclusions (see text for explanation).

that the smooth pursuit system is either defective or totally inoperable.

The second observation derived from Fig. 3 involves the indiscriminate use of the slow phase eye velocity to determine smooth pursuit gain when a spontaneous nystagmus was present during fixation of a stationary target. Attempts to do so will cause gains greater than 1.0 for tracking in the direction of the slow phase and either less than 1.0 ('low gain'), 0.0 ('no pursuit') or negative gains (so-called 'reversed pursuit') for tracking in the direction opposite to the slow phases; the relative velocities of the target and spontaneous slow phases will determine which of these erroneous interpretations results. It is the last case, where the nystagmus slow phases are at higher velocities and in the opposite direction to the target, that is most common in CN due to large shifts in the neutral zone from the SNZ. When this is combined with the usual practice of measuring pursuit only in the central 40° of gaze, the pursuit does not extend through the dynamic neutral zone (DNZ). If it did, CN direction reversal would be observed at some point in the record and the mistaken interpretation of 'reversed pursuit' discredited. Again, simply stated, in the presence of spontaneous nystagmus, slow phase velocities are *not* equal to

pursuit velocities and cannot be used to calculate pursuit gain; the slow phase velocities of the spontaneous nystagmus must first be removed.

When examining the effects of an ongoing nystagmus on the smooth pursuit response of a normal, we found responses like those shown in Fig. 3 (Abel *et al.*, 1979; Daroff *et al.*, 1979). Although the nystagmus was induced by constant, cold-water canal irrigation in a subject with perfectly normal smooth pursuit, he could not generate *any* pursuit eye movements in the direction of the target motion and opposite to the slow phases of the caloric nystagmus. In that case we knew that smooth pursuit was normal but had we applied the same criterion that is commonly used when testing patients with spontaneous nystagmus, we would have been forced to conclude that this subject had no smooth pursuit. As was the case in so-called 'pursuit-defect' nystagmus, it was the expectation of the investigators that was defective not the smooth pursuit of the patient. Just as a normal could not override the low velocities of caloric nystagmus with smooth pursuit, neither can the CN patient override the much faster slow phases of his nystagmus with his equally normal smooth pursuit.

If we are to evaluate properly the function of the smooth pursuit subsystem or calculate smooth pursuit gain in a CN patient by the same methods used in normals, we must use patients who have substantial foveation periods in their waveforms or large null zones containing virtually no nystagmus. Only during these periods can the eye velocity equal target velocity and the gain be calculated by the ratio of the two. At all other times, the best method is to linearly interpolate between the foveation periods of the CN waveforms (no matter how short they are) and calculate the smooth pursuit gain using the slope of the resulting curve. This, in effect, subtracts out the ongoing CN waveforms and is also the method of choice in acquired nystagmus where flat foveation periods are not likely. In rare CN cases, we may find smooth pursuit accomplished by the matching of slow phase velocity to target velocity; this requires deceleration as well as velocity matching of the accelerating slow phases and is, therefore, a difficult task for the ocular motor system. Since there are no runaway slow phases taking the eyes off target, there will be no corrective fast phases (*i.e.*, the nystagmus will be suppressed).

PURSUIT DATA FROM CN PATIENTS

The data collected from the first CN patient to be quantitatively studied included pursuit of a sinusoidally moving target and from a target moving with constant velocity (Dell'Osso, January 1968; Dell'Osso *et al.*, 1972). Various sinusoidal frequencies and constant velocity movements were used. Gain and phase plots of the CN patient's pursuit were found to be equivalent to those of normals. The figures and discussion above are a direct result of the initial hypothesis, embodied in the model (Dell'Osso, January 1968), that the smooth pursuit system of a CN patient is normal and the pursuit eye movements are the result of the superposition of the output of that normal system and the ongoing (and changing) CN. The following data were selected from studies of over 400 patients made over the past 15 years. They illustrate the points made above regarding the expected eye movements from patients with spontaneous nystagmus.

When evaluating pursuit data, the desire to find long periods of uninterrupted smooth pursuit should be tempered with the realization that any given patient may have a perfectly normal smooth pursuit system and still not generate *any*

substantial periods of smooth eye movements. The eye movements generated will depend on the patient's CN waveforms (there are usually several) and how they change during the attempt to pursue (*i.e.*, how much null shift is present in each direction). The method of joining foveation periods and analyzing the slope of the resulting curve is the only *reliable* test of the smooth pursuit system in the presence of CN. Because of the importance of recognizing the foveation periods of each of the 12 CN waveforms and of the transitions between waveforms, it will be helpful to have some aid in identifying these periods and thereby tracing the line of foveal fixation. The identification of foveation periods for all CN waveforms has been extensively discussed elsewhere (Dell'Osso & Daroff, 1975). On various segments of the records shown below I have indicated those periods corresponding to target foveation (or tracking) to clarify the accompanying descriptions. These periods are easy to see for some waveforms but are particularly difficult during transitions from one waveform to another. Due to waveform variation, not all parts of every record show accurate pursuit; neither CN patients nor normals (over-achieving graduate students excepted) consistently follow moving, featureless spots of light perfectly (gain = 1.0) and our aim, when evaluating smooth pursuit of either normals or patients, has always been to demonstrate their ability to pursue accurately *when they chose to do so.* We are interested in establishing the viability of the pursuit system and measuring its gain under the best of conditions. Too many factors can interfere with good smooth pursuit and yield false impressions of partial or complete failure of pursuit; boredom and attention lapses (easily induced by most laboratory tasks) are especially deleterious to good pursuit.

Figs. 4-9 contain data taken on curvilinear paper in the early 1970's before we converted to rectilinear paper; for the analysis of smooth pursuit near primary position the effects of curvilinear paper are minimal. Fig. 4 shows pursuit of a CN patient who had jerk with extended foveation (J_{ef}) and pseudo-pendular with foveating saccades (PP_{fs}) superimposed upon his smooth pursuit. The nystagmus during leftward pursuit is always jerk left since the DNZ has been shifted far to the right. The nystagmus during rightward pursuit varies between jerk left, in far left gaze, and PP_{fs}, from near left gaze throughout right gaze. As indicated, while pursuing to the right the DNZ extends from 3° left to 10° right. The velocity traces show the DNZs most clearly since the large velocity spikes will be of opposite directions on either side of a DNZ and, as in this case, the alternating direction of the velocity spikes of bidirectional jerk waveforms will define the extent of the DNZ. The lines joining foveation periods show linear smooth pursuit to the right (with an initial catch-up saccade) and to the left. The gains, calculated using these lines of foveal pursuit for this 4°/sec target motion, were .96 and 1.0 for rightward and leftward pursuit, respectively. Note the three spontaneous bias reversals during the first instance of leftward pursuit (at arrows); such bias reversals occur many times during steady fixation as well. They represent sporadic reversals of the instability causing the slow phase to go off in the wrong direction and are unrelated to the smooth pursuit. In the push-pull neural integrator model, bias reversals correspond to the opposite integrator going unstable in a region where it is normally stable.

Fig. 5 shows the same patient pursuing a target moving sinusoidally with a continuously decreasing frequency. The modulation of the position of the CN oscillation is clear. The

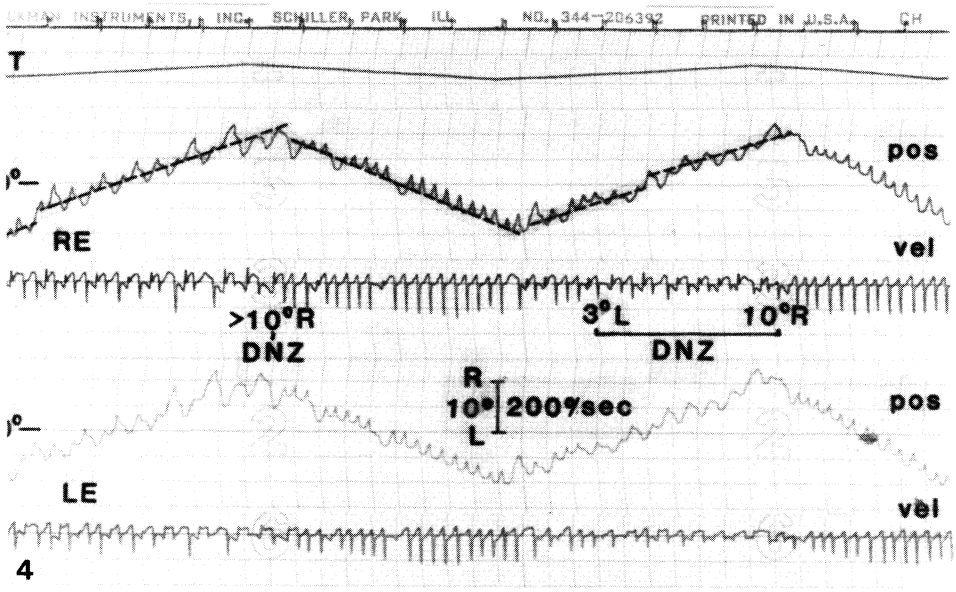

Fig. 4. Example of a CN patient pursuing a target moving with a constant velocity (ramp) of 4°/sec. In this and the following figures: RE = right eye; LE = left eye; R = right and L = left directions; pos = eye position; vel = eye velocity; T = target position; DNZ = dynamic neutral zone; timing marks are at 1 second intervals; and dashed lines (when shown) indicate the patient's line of sight and are constructed by joining the foveation periods of each CN beat; this is equivalent to subtracting out the ongoing and changing CN. In Figs. 4-8, thickness of the dashed lines corresponds to a visual angle of 20 minarc; the 1° foveal area is, therefore, equal to three times that thickness. Note the DNZ shift opposite to pursuit direction. Target not shown to scale in this figure; excursion is ± 10°.

dashed curve over the right eye record is a tracing of the target motion fitted over the eye movements. The DNZs are indicated for pursuit in both directions for the pursuit of the more slowly moving portions of the record. During the more rapid pursuit the DNZs have shifted too far to be seen within the ± 10° range of target motion and pursuit to the right contains only jerk right CN (DNZ beyond 10° left) and pursuit to the left, jerk left (DNZ beyond 10° right).

Fig. 6 is the pursuit of another patient who shows a large region of PP_{fs} with only a few pseudo-jerk right (PJR) beats while pursuing leftward. Reflecting the shifts opposite to pursuit direction, the DNZ extends over the whole 20° range of rightward pursuit and from 10° right to 0° during leftward pursuit. The beginning of the PJR waveform defines the end of the DNZ since PJR is actually a jerk left CN (see velocity trace for direction of velocity spikes). The rightward and leftward pursuit gains, calculated using the lines of foveal pursuit for this 4°/sec target motion, were .91 and .96, respectively.

Fig. 7 illustrates the ability of another patient to pursue by suppression of slow phase acceleration. The CN remains leftward J_{ef} during pursuit in both directions, indicating that the DNZ is beyond 15° to right for leftward pursuit but remained at 5° right (the static null) for rightward pursuit at this low velocity (12°/sec). The

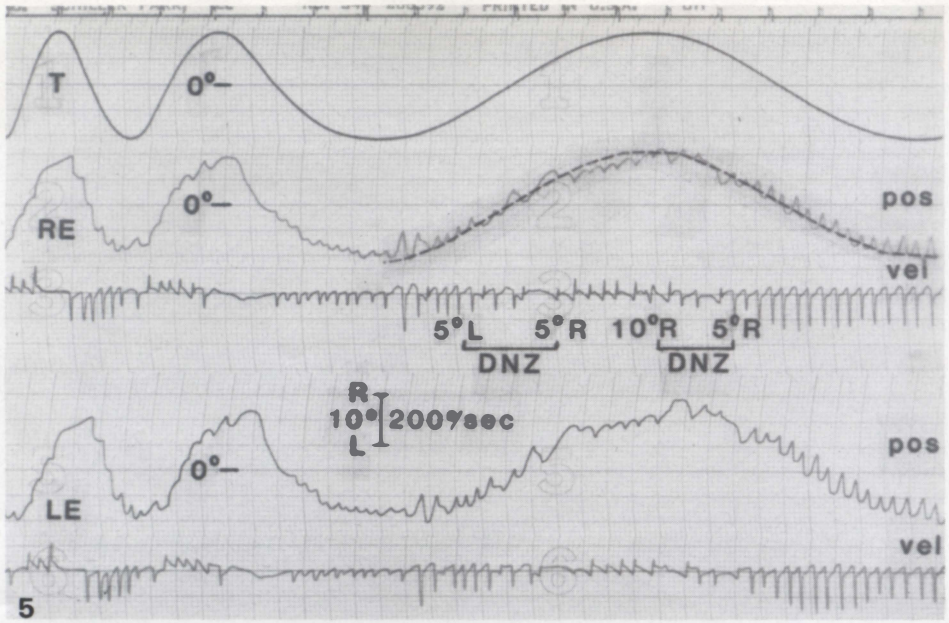

Fig. 5. Example of a CN patient pursuing a target moving sinusoidally with decreasing frequency. Dashed curve over right eye record is target motion placed over patient's eye movements. Note the DNZ shift opposite to pursuit direction. Scale applies to T also.

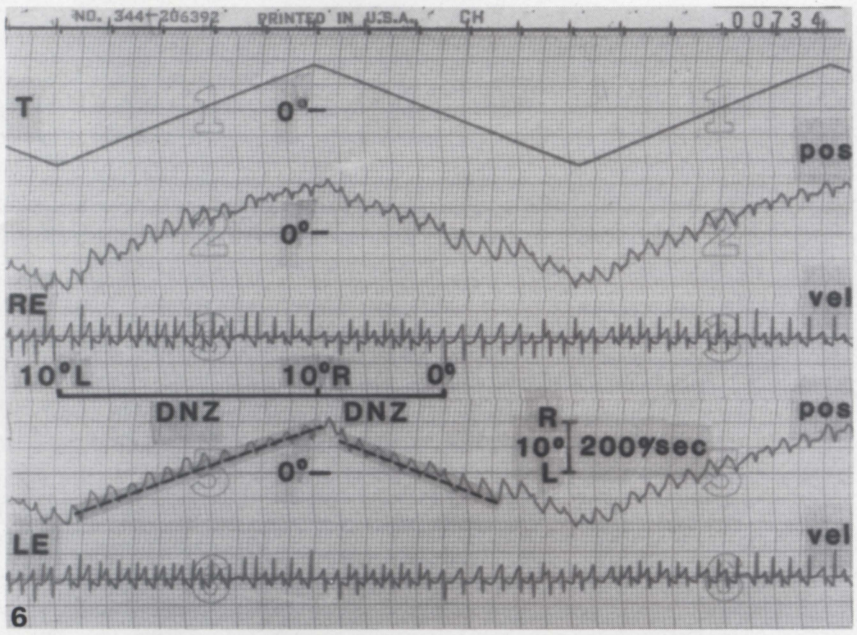

Fig. 6. Example of a CN patient pursuing a target moving with a constant velocity of 4°/sec that illustrates large dynamic neutral zones. Note the DNZ shift opposite to pursuit direction. Scale applies to T also.

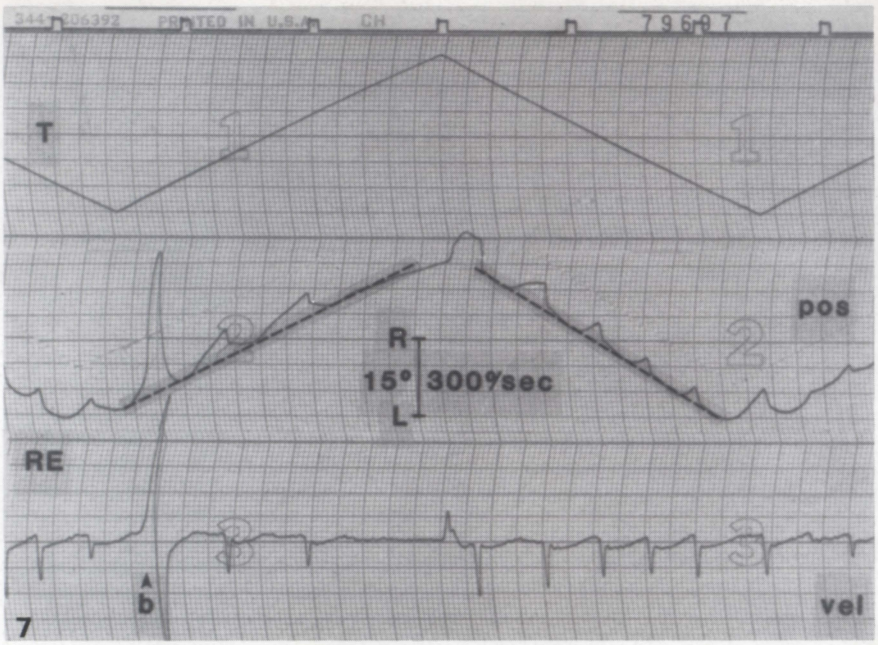

Fig. 7. Example of a CN patient pursuing a target moving with a constant velocity of 12°/sec that illustrates pursuit by suppression of slow phase acceleration. Scale applies to T also.

normally flat (*i.e.*, zero velocity) foveation periods show pursuit to the left and when pursuing to the right, the accelerating slow phase is actually slowed in an attempt to match the target velocity. The rightward and leftward pursuit gains, calculated using the lines of foveal pursuit, were 1.04 and 1.19, respectively.

Fig. 8 shows smooth pursuit to the left and right of the same patient at a faster target velocity (30°/sec) with total CN suppression when pursuing to the right. The DNZ is now beyond 15° left for rightward pursuit and further to the right than in Fig. 7 for leftward pursuit. The rightward pursuit gains varied from .57 to 1.25. The leftward pursuit gain, calculated using the lines of foveal pursuit, was .80. If one looked at only the rightward pursuit, it could not be identified as pursuit of a CN patient; the only saccades present are the normal initial catch-up saccades in the first and third sections plus an additional one in the third. Note that by anticipating this predictable target motion in the second section, the patient was able to use a slow phase to generate a smooth pursuit segment without an initial saccade. These three sections of rightward pursuit duplicate the three most common pursuit responses of normals.

The final figure from the early 1970's demonstrates another normal, albeit rare, ability to use the pursuit system. Fig. 9 shows the same patient as in Fig. 6 *imagining* a slowly moving target moving left and right in the dark. Some imagined rightward pursuit is evident during the foveation periods (compare them with the flat foveation periods of steady fixation in the first two beats) but it was difficult for the patient to

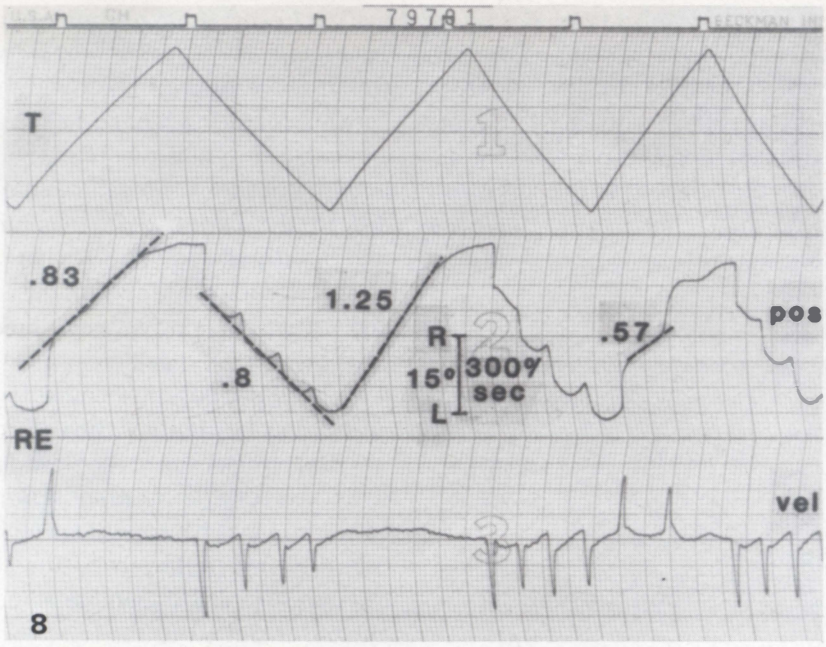

Fig. 8. Example of a CN patient pursuing a target moving with a constant velocity of 30°/sec that illustrates total suppression of slow phase acceleration when pursuing a higher velocity target. Scale applies to T also.

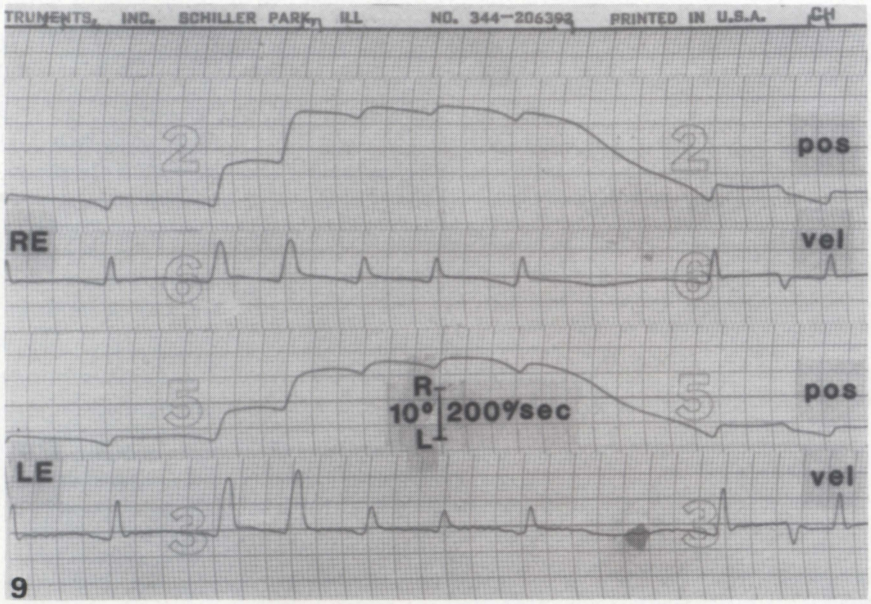

Fig. 9. Example of a CN patient pursuing an *imaginary* target moving with constant velocity that illustrates suppression of slow phase acceleration during pursuit of a mental image of the target.

overcome the strong leftward slow phases of his jerk right nystagmus. When pursuing to the left, however, he utilized the leftward slow phases to generate smooth movements to the left (note that to do this he had to *halt and reverse* the acceleration of the slow phases to the left also). Without a moving target, not many normals can generate smooth pursuit.

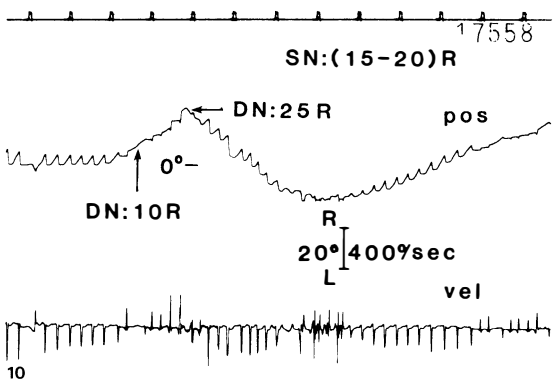

Fig. 10. Example of a CN patient pursuing a hand-held target to demonstrate the large dynamic null shifts induced by pursuit. The centers of the neutral zones are indicated in degrees for the static null (SN) and dynamic nulls (DN).

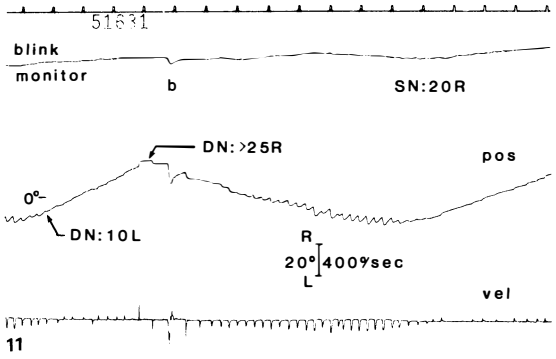

Fig. 11. Example of a CN patient pursuing a hand-held target to demonstrate the large dynamic null shifts induced by pursuit. The centers of the neutral zones are indicated in degrees for the static null (SN) and dynamic nulls (DN).

Figs. 10 and 11 are recordings of a more modern vintage (1977) using infrared reflection and rectilinear recording. The figures show two CN patients following a hand-held target light and are included to illustrate the dynamic null shifts in both directions; no gains could be calculated due to the nature of the stimulus. In these figures the null angles (the centers of the neutral zones) are indicated for both the static null (SN) and, when pursuing in each direction, the dynamic nulls (DN) (Dell'Osso, 1984). Fig. 10 contains good periods of extended foveation before pursuit has begun, including the one spontaneous bias reversal where the slow phase accelerated in the wrong (leftward) direction. The SN was between 15 and 20° right. During rightward pursuit, it shifted to the left by 5 to 10° so that the DN appeared at 10° right gaze; during leftward pursuit, it shifted to the right so that it was at 25° right gaze. Smooth pursuit is apparent in both directions during the foveation periods. Fig. 11 shows another patient whose SN was at 20° right gaze but whose null during pursuit to the right moved 30° to the left; the resulting DN appeared at 10° left gaze. During pursuit to the left, the null moved more than 5° to the right, placing the DN beyond 25° to the right. This patient had a poor waveform with little foveation time per cycle and yet there is good pursuit to the right when the patient passed through the null zone where the CN is minimal; we should not expect to see it during this sawtooth-like waveform. The effects of his smooth pursuit to the left, on the rightward slow phases, are most evident in right gaze near the null and become less effective as he pursues further away from the null where the more rapidly accelerating slow phases predominate.

The final four figures show data collected in 1985 using infrared reflection and rectilinear recording. Fig. 12 illustrates the long periods of

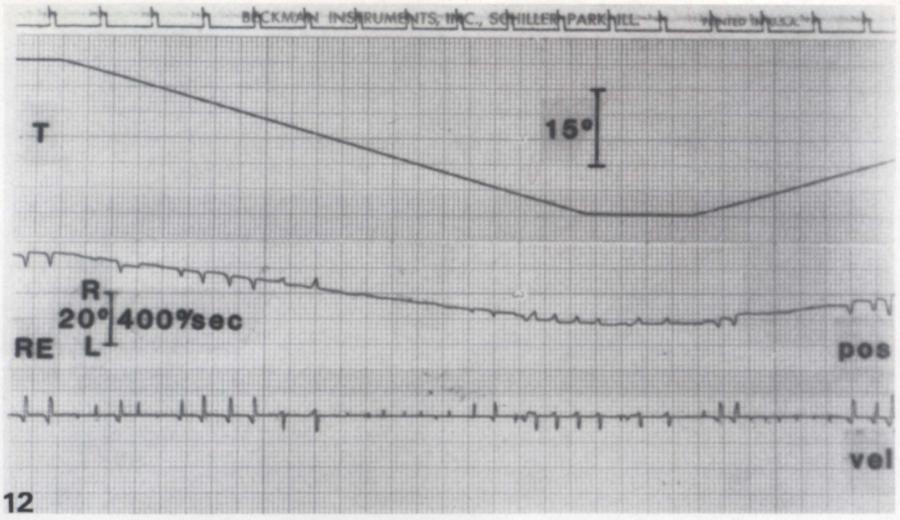

Fig. 12. Example of a CN patient pursuing a target moving with a constant velocity of 3°/sec that illustrates long periods of foveal pursuit during the extended foveation periods as well as spontaneous bias reversals during unidirectional (leftward) pursuit.

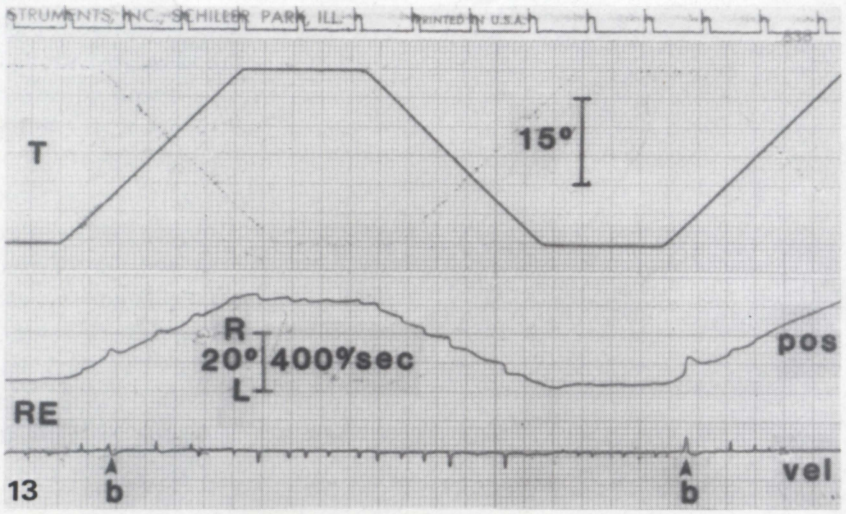

Fig. 13. Example of a CN patient pursuing a target moving with a constant velocity of 10°/sec that illustrates pursuit during foveation periods.

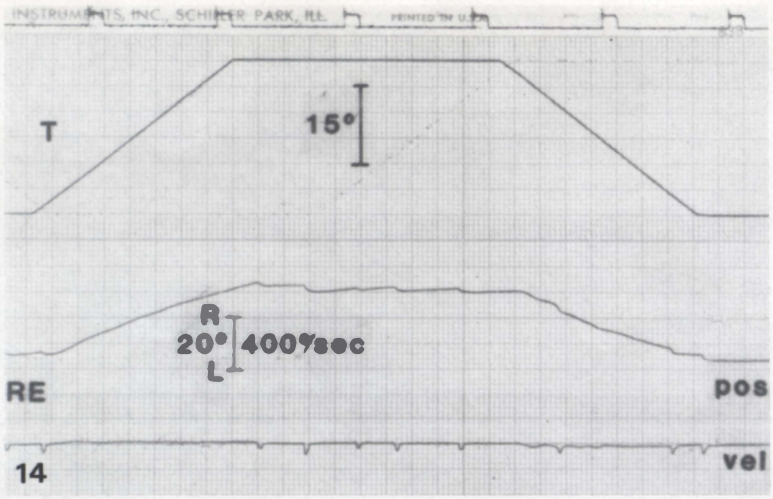

Fig. 14. Example of a CN patient pursuing a target moving with a constant velocity of 20°/sec that illustrates suppression of the CN in one direction (rightward).

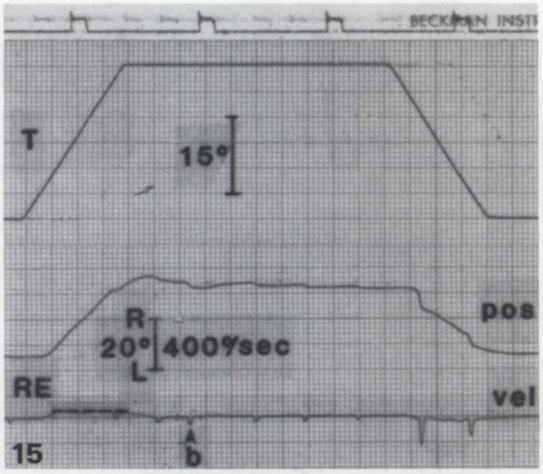

Fig. 15. Example of a CN patient pursuing a target moving with a constant velocity of 40°/sec that illustrates suppression of the CN in both directions. Note constant velocity tracking (dashed line) in velocity tracing.

good smooth pursuit possible in patients whose waveforms have substantial extended foveation periods. When the target was stationary, the eye velocity was zero during these periods and during smooth pursuit at 3°/sec, it was equal to target velocity. Thus, the smooth pursuit gain was equal to 1 during target foveation. This occurred *independently of the slow phase direction* as can be seen in the pursuit to the left where several spontaneous reversals caused the slow phases to accelerate to the right. In cases such as this, both the method of joining foveation periods to calculate pursuit gain and that of averaging the individual gains calculated for each foveation period will yield equivalent values.

The data in Figs. 13-15 are from another patient and illustrate the effects of target velocity on the CN and on the patient's ability to suppress the CN entirely and generate only his smooth pursuit response. The target velocity in Fig. 13 was 10°/sec and tracking to the right consisted of unity-gain foveation periods. Since

the target could only be tracked to the left during short foveation periods due to the strong rightward slow phases (the DNZ was far to the right), the gain was calculated by the method of joining these periods; it also had a value of 1. In Fig. 14, where the target speed was 20°/sec, the patient was able to suppress the CN and pursue fairly smoothly in both directions; the rightward gain varied from .90 to 1.10 and the leftward gain from .70 to 1.10. At 40°/sec, in Fig. 15, the CN was fully suppressed and pursuit in both directions was indistinguishable from that of a normal. The gains were approximately equal to 1 for rightward pursuit and .88 for leftward pursuit.

CONCLUSIONS

The study of fixation and smooth pursuit of CN patients has resulted in more general definitions of 'normal' functioning of these two subsystems. They are equally applicable to both normals and patients with ocular oscillations whereas the commonly used definitions are very limited in their application and can be misleading under all but the most contrived circumstances. The new, broader definitions are derived from, and consistent with, current conceptualizations of ocular motor control. It is possible to have a normally functioning fixation reflex despite the presence of spontaneous oscillations and it is equally possible to have a normal smooth pursuit system operating in the presence of these same oscillations. It is my contention that these are the conditions that prevail in most neurological and congenital ocular motor oscillations; they are certainly the conditions present in CN patients.

The data contained in this report spans 15 years. However, most of the observations made in support of my original hypothesis, that normal pursuit was superimposed on the changing CN waveforms, are evident from the data contained in Figs. 4-9 which were obtained in 1972 and 1973 from only the second, third and fourth CN patients we studied. Subsequent, and more accurate, recordings of more than 400 patients have continued to support this hypothesis and represent a huge body of data whose existence refutes recent suggestions that patients with CN have poor or absent smooth pursuit (Yamazaki, 1979; Yee *et al.*, 1981; Kommerell & Mehdorn, 1982; St John *et al.*, 1984; Collewijn *et al.*, 1985) or can only generate 'reversed' pursuit (Optican *et al.*, 1983). Many of these papers have been discussed elsewhere (Dell'Osso, 1982a, 1984), as have some of the concepts contained herein (Dell'Osso, 1982b). Therefore, it is neither necessary nor desirable to present detailed rebuttals to such contentions; they can all be traced to the failure to consider the points raised above. Furthermore, the *data* contained in some actually support the hypothesis of good smooth pursuit added to prevailing CN (see below); one has merely to evaluate the eye movements properly. The spontaneous bias reversals shown in Fig. 12 during leftward pursuit are a direct contradiction to the claim that reversed pursuit is the cause of CN. In this record we see slow phases in *both* directions while smooth pursuit to the left is unchanged during the extended foveation periods. Thus, the mechanism dictating the CN slow phase direction is independent from that generating the pursuit signal.

Despite statements in some papers that suggest otherwise, they contain eye movement data that support the hypothesis of intact smooth pursuit in CN patients (Halmagyi *et al.*, 1980; Kommerell & Mehdorn, 1982; Larmande *et al.*, 1983). In Fig. 4 of Halmagyi *et al.* (1980) the DNZ pursuit-induced shift was illustrated. The SNZ extended from 5° left to 15° right. During

rightward pursuit the CN was all jerk left, indicating a DNZ far to the right (greater than the 25° rightward extent of the target motion) and during leftward pursuit the CN was all jerk right, indicating a DNZ far to the left (again, beyond the 25° leftward extent of target motion). My contention that target foveation is accomplished at the same point in the CN waveform during pursuit as it is during fixation was shown by the alignment of those foveation periods just before and just after the target stopped; this despite the obligatory reversal of the CN caused by the DNZ moving back to the SNZ. In their Fig. 4, Kommerell & Mehdorn (1982) demonstrated both the DNZ shifts with pursuit and their increase with pursuit velocity. In Fig. 4A slow pursuit resulted in right pseudocycloid (RPC) waveforms for both leftward and most of rightward pursuit. During leftward pursuit the DNZ was beyond the 20° extent of target motion but during rightward pursuit it shifted to the region between 0° and 5° left where the CN was pseudopendular with foveating saccades. In Fig. 4B faster pursuit resulted in greater DNZ shifts so that the waveforms were RPC and LPC during rightward and leftward pursuit respectively (*i.e.*, DNZ shifts were leftward and rightward respectively). Thus, the DNZ that had shifted rightward to 0° during slow left pursuit, shifted further to the right to beyond 20° right during fast leftward pursuit. In Fig. 2 from Larmande *et al.* (1983) there was, in addition to DNZ shifts, a clear example of pendular CN added to the ramp of smooth pursuit to the right during pursuit through the DNZ. Since there were no saccades present, the only way to produce such eye movements is by adding good smooth pursuit to the pendular waveform. Although I disagree with some of the conclusions drawn in the above papers, the data speak for themselves.

In addition to the above papers, which contain data supporting my contention that CN patients can pursue normally, there are other studies where, by correctly interpreting their data, the authors have reached the same conclusion (Gresty *et al.*, 1984; Kommerell, 1986). Halmagyi *et al.* (1980) and Gresty *et al.* (1984) have attributed the appearance of 'reversed' pursuit and 'reversed' OKN to null shifts that actually reverse the CN. Kommerell (1986) has shown that CN patients, like normals, can generate slow eye movements in the direction of a parafoveal after-image and has conceded that they can indeed generate smooth pursuit but not by the retinal slip velocities that they too can perceive. Citing only evidence equally exhibited by normals, he has hypothesized that the pursuit of CN patients is due solely to position information and not the retinal slip he has shown they can perceive. How then do they pursue during the flat foveation periods where there is no position error and yet the eye velocity continues to match that of the target? Also difficult to reconcile are the statement that CN is due to 'a primary defect of retinal slip control' and the observation that 'timing and direction of slow and rapid components of CN do not depend on *retinal* feedback' (the latter has been known for some time (Dell'Osso *et al.*, 1974)).

We have demonstrated that by identifying the DNZ in records of pursuit in each direction a shift in its position opposite to pursuit direction will be seen. When does this shift occur? From the observation that the CN waveforms changed abruptly when pursuit direction changed as a patient pursued a target moving in alternate directions with constant velocity (*i.e.*, a triangular wave as in Figs. 4 and 6-8), we had previously inferred that the DNZ shift occurred with the onset of pursuit. Optican *et al.* (1983) studied the responses of CN patients to step-ramp

stimuli. The significant, but unrealized, finding of these authors was that their paradigm proved the pursuit-induced neutral zone shift in CN is instantaneous and simultaneous with the onset of smooth pursuit. The fact that the CN of one of their patients reversed 130 msec after the onset of the stimulus showed that the DNZ was already shifted at the same time that he began to pursue (accepting 130 msec as the latency for smooth pursuit). Their paradigm also confirmed previous observations that CN does not depend on retinal slip; fixation attempt had been shown to be responsible for the genesis and intensification of CN independent of either retinal illumination or lid position (Dell'Osso, 1973; Dell'Osso *et al.*, 1974). The Rashbass stimulus proves that normal smooth pursuit responds to retinal slip velocity since it is in a direction opposite to the initial step of target position. Similarly, the response of a CN patient to this novel stimulus also proves that this pursuit system responds to the slip signal and not the position signal. The CN reversal was in the direction dictated by pursuit in the proper direction and not towards the new target position; this observation supports the hypothesis of normal pursuit in response to target velocity and disproves the contention that pursuit in CN is carried out in response to position error (Kommerell, 1986).

One of the dangers of misidentifying CN slow phases as reversed smooth pursuit is evident in the report of a model for CN that relies heavily on a reversed velocity feedback loop to generate CN-like waveforms (Optican & Zee, 1984). A thorough discussion of this complex model is beyond the scope of this paper. The model successfully suggested and examined mechanisms that might be responsible for the genesis of CN; smooth pursuit was not discussed. However, since the model was based on reversed smooth pursuit, it is difficult to conceive of any way that it could produce the good smooth pursuit illustrated in both the figures contained herein and those in the references listed above.

Since many patients with pendular CN also have jerk CN and since the genesis for both is similar, the early data showing smooth pursuit superimposed on a pendular waveform was sufficient to establish intact smooth pursuit in CN patients whatever their waveform. This is evident in patients whose waveforms change during smooth pursuit from jerk to pendular or *vica versa.* For the limited target excursions and velocities normally used to test smooth pursuit in the laboratory (± 20° and less than 40°/sec, respectively), smooth pursuit is a weak stimulus with limited velocity capabilities and cannot be expected to override the slow phases of ongoing nystagmus. We demonstrated this with normals given induced caloric nystagmus who were unable to pursue targets moving in a direction opposite to the slow phases of the caloric nystagmus. Obviously, we could not conclude that such subjects had impaired smooth pursuit since they were known *a priori* to be normal. Given the greater intensity of the CN slow phases when compared to caloric nystagmus, it is curious that some still expect to see smooth pursuit overriding slow phases that reach velocities over 200°/sec; that is clearly an impossible feat for most smooth pursuit systems and it is, therefore, impressive when some CN patients actually accomplish it.

The first priority of the brain of a CN patient is most probably the prevention of oscillopsia due to the constantly moving retinal images. I have hypothesized that this is accomplished by utilizing normally-present efference copy to negate the nystagmus-induced retinal image motion present in the light (Dell'Osso, 1967; January 1968). In this straightforward way, both

the saccadic and pursuit systems remain uncompromised by additional responsibilities and can function normally using the resulting true retinal position and velocity error signals. The CN patient has no trouble perceiving such changes in target position or velocity despite his oscillation (Kommerell *et al.*, 1986).

Is it reasonable to suggest that a normal smooth pursuit system would not tolerate the retinal slip velocities caused by CN? Supposedly, according to this line of reasoning, they would stimulate the pursuit system and it would stabilize the eyes (this implies that neither CN nor any other nystagmus should be possible if the pursuit system were normal). Ignoring for the moment the fact that vestibular nystagmus can easily be induced in normals *despite their intact pursuit systems*, such a conjecture fails to conform to the basic nature of smooth pursuit: to enable the *willful* pursuit of a small target, with *real, perceived* or *imagined* motion, across a featured background. The smooth pursuit system does *not* function to stabilize all moving retinal images; on the contrary, it must ignore them when they constitute the background. In the case of a CN patient fixating a stationary target, there is no willful pursuit nor is there real, perceived or imagined target motion and, therefore, the pursuit system can neither be activated nor used to cancel the CN. Even if there were a way to use pursuit for that purpose, it would probably be counterproductive since such use could interfere with or prevent true pursuit of a moving target. By utilizing the available efference copy to enable perception of a stable world, the remaining ocular motor subsystems are left free to accomplish the specific tasks for which they evolved; my contention that they do so is supported both by the data and the absence of any symptoms of malfunction in the saccadic, pursuit or vestibular systems.

There is ample evidence that areas in the temporal cortex have access to both retinotopic (area MT) and target (area MST) velocities (Newsome *et al.*, 1985; Wurtz & Newsome, 1985; Sakata *et al.*, 1983). Yasui & Young (1975) have shown that efference copy is used by the ocular motor system to reconstruct a target velocity signal that is used to drive the smooth pursuit system. Thus, all the required signals and pathways needed by the CN patient to both stabilize the world and pursue targets of interest are present in normals; this obviates the need to postulate exotic neural connections. The only prerequisite for their use seems to be that the nystagmus be present during early developmental years since adults with acquired nystagmus cannot overcome the oscillopsia caused by their oscillations. Our preliminary studies on oscillopsia and CN, using stabilized retinal images, suggest that image motion *per se* is not the key variable. It appears that mental set determines whether a target is 'moving' or 'stationary' with respect to the background. Thus, once the CN-induced retinal motion is perceived as 'stationary' by the brain, any other retinal motion is correctly perceived as target motion and can be pursued at will.

The model in Fig. 16 is an updated and slightly modified version of my original hypothesis (Dell'Osso, 1967; January 1968). The saccadic system and plant dynamics have been removed for simplicity and the original pursuit system replaced by one suggested by the work of Yasui & Young (1975). The internal perception of world velocity is given by:

$$\dot{W}' = \dot{W}/[1 + (1-K)G], \text{ where } G = G_{ol}.$$

During fixation of a stationary target ($\dot{W} = 0$), $\dot{W}'$ will also equal zero and there will be no oscillopsia, independent of the values of K or G

(these are 0.7 and 1.4 respectively, in normals). At all times, $\dot{E} = \dot{E}' + \dot{N}$ (*i.e.,* eye velocity equals the eye velocity command plus the nystagmus velocity). In light, the value of eye velocity $\dot{E} = \dot{N} + G\dot{W}/[1 + (1-K)G]$ which, during fixation on a stationary target ($\dot{W} = 0$), reduces to $\dot{N}$, independent of K or G. The switch 'S' is included since this pathway is utilized to prevent oscillopsia and none is possible in the dark. Therefore, in the dark, $\dot{E} = \dot{N}$ where the value of $\dot{N}$ is dependent on 'fixation attempt' or 'effort to see' as has been documented elsewhere (Dell' Osso, 1973; Dell'Osso *et al.,* 1974). Pertinent to this paper, the model predicts that during smooth pursuit of a moving target with velocity $\dot{W} = \dot{T}$, $\dot{E} = \dot{N} + G\dot{T}/[1 + (1-K)G]$. For the normal values of K and G given above, this results in eye velocity $\dot{E} = 0.98\dot{T} + \dot{N}$, where the eye velocity command $\dot{E}' = 0.98\dot{T}$. Thus, when $\dot{N} = 0$ (during foveation periods), $\dot{E} = \dot{E}' \cong \dot{T}$ (*i.e.,* the eye velocity command is approximately equal to the target velocity, indicating normal smooth pursuit). At all times, the model predicts normal smooth pursuit superimposed upon the existing nystagmus.

In the 20 years since the original hypothesis of normal smooth pursuit superimposed on the CN waveform was advanced in the form of a model similar to that in Fig. 16, much has been learned about waveforms, fixation, nulls and other characteristics of CN. Better recording techniques have also yielded more details about the eye movements of CN patients. Supported by all the new information that has resulted, it is my opinion that there has emerged no better explanation for the operation of the various ocular motor subsystems than that embodied in the above model. It is consistent with all of the new data on CN as well as with our increased understanding of the ocular motor mechanisms responsible for moving the eyes. None of the recent hypotheses that have suggested poor or 'reversed' pursuit in CN patients have included an explanation of which of the ocular motor subsystems could possibly be responsible for the periods of eye motion where position and velocity match those of the target on a beat-to-beat basis. If not smooth pursuit, what is the mechanism responsible? I submit that only an intact smooth pursuit subsystem could accomplish the normal pursuit evident in the figures above and that more will be learned about smooth pursuit when we ask the right questions about how the CN patient harnesses his pursuit system to do the job than has resulted from protestations that something *must* be wrong with it. There is a wealth of evidence in support of good pursuit and a dearth of symptoms that might suggest its absence. It is time to return to the study of what the pursuit system *does* under normal and abnormal circumstances rather than continue with unfounded speculation on what it *should* be able to do, given those circumstances.

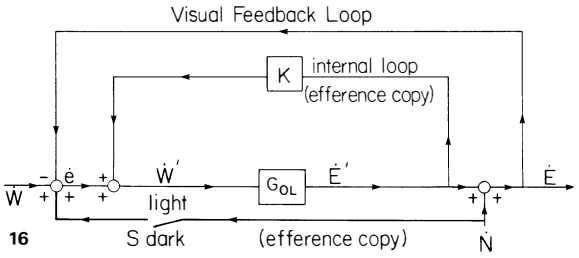

Fig. 16. Model of smooth pursuit system and congenital nystagmus (Ṅ). Ẇ is world velocity, ė is retinal error (slip) velocity, Ẇ′ is reconstructed world velocity, G_{ol} is open loop gain of the pursuit system, K is gain of the positive feedback internal loop of the pursuit system, Ė′ is eye velocity command and Ė is eye velocity. The switch S allows use of the feedback signal Ṅ to prevent oscillopsia in the light. The first summing junction is a lumped summing point and is not meant to imply that the efference copy of the nystagmus goes to the retina. Rather, it is used cortically to produce a retinal slip velocity that is independent of the nystagmus.

In addition to the claims of poor smooth pursuit, the literature contains equally doubtful statements about the vestibulo-ocular response (VOR) of CN patients (Yamazaki, 1979; Yee *et al.*, 1981; Carl *et al.*, 1985; Gresty *et al.*, 1985; Demer & Zee, 1984) and the optokinetic nystagmus (OKN) system (St John *et al.*, 1984; Yee *et al.*, 1980; Kommerell & Mehdorn, 1982; Collewijn *et al.*, 1985; Halmagyi *et al.*, 1980; Larmande *et al.*, 1983; LeLiever & Barber, 1981; Abadi *et al.*, 1982; Collewijn *et al.*, 1978). Without going into detail, it should be obvious that the above considerations concerning the analysis of pursuit in the presence of CN apply equally to calculating the gains of the VOR or OKN systems; the existing CN slow phases must be subtracted from the resulting slow eye movements first and then the gains determined. Our data indicate normal VOR, VOR suppression and circularvection induced by OKN stimuli. Because of the need to prevent oscillopsia, it is probable that the CN patient adapted by suppressing his OKN response somewhat. However, the OKN stimulus is having the same effect on the CN as pursuit since it does cause a DNZ shift and CN reversal. Other authors support the contention of good VOR and OKN-induced CN reversal rather than the erroneous idea that the OKN response is somewhat reversed (Gresty *et al.*, 1984; Abadi & Dickinson, 1985; Abadi *et al.*, 1982).

ACKNOWLEDGEMENT

This work was supported in part by the Veterans Administration.

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