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CONGENITAL NYSTAGMUS WAVEFORMS AND FOVEATION STRATEGY

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ABSTRACT

Accurate eye movement recordings of sixty-five subjects with congenital nystagmus (CN) provides a firm foundation for the classification of the many types of waveforms observed and results in objective definitions based on measurable quantities rather than subjective clinical impressions. The careful scrutiny of these records along with the utilization of laser-target retinal cinematography have yielded insights into the mechanism of this ocular motor instability.

Prolongation of target foveation emerges as the dominant factor in many of the resultant waveforms. This enhances the visual acuity of the subject with CN. An additional observation, related to fixation bias reversals of the CN subject, may be a physiological indicator of foveal function.

Utilization of accurate, DC-coupled eye movement recording techniques in the study of congenital nystagmus (CN) has resulted in the use of composite prisms to improve visual acuity of patients with this disorder (Dell'Osso et al., 1972; Dell'Osso, 1973; Dell'Osso et al., 1974). The quantitative recordings and the use of retinal cinematography have provided insights into the genesis of this ocular motor instability and a sounder basis for waveform classification. In this report we will identify previously held beliefs about CN which we have found to be erroneous, and will re-define and classify the nystagmus types and waveforms in a manner based on measurable quantities which are free from confusing and oft-times misleading clinical impressions. Whenever possible, we will relate waveform to the foveation strategy utilized to enhance visual acuity.

METHODS

The eye movement recording and the laser-target retinal cinematographic techniques have been described in detail in the previously cited publications and will be reported only briefly here. Eye movements were recorded from each eye individually with electro-oculography or an infrared reflection system. The electronics employed were DC-coupled with a bandwidth of

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25 Hz or 100 Hz, depending on the accuracy required for a particular recording. Retinal cinematography was accomplished using a 50μ laser target imaged onto the retina through a −50D contact lens. Using the same optics, the retina was illuminated with white light and the foveal motion recorded with a 16 mm camera. Over 65 subjects (44M and 21F) have been studied, ranging in age from 33 to 67 years.

RESULTS

The major conclusions of our previously published studies will be presented initially and contrasted with commonly stated misconceptions about congenital nystagmus.

Pendular (P) nystagmus is a sinusoidal-like oscillation of the eyes biased such that the fovea rests on the target at one or the other peak of the waveform (Dell'Osso, 1973). Thus, descriptions of pendular CN which describe two-and-fro movements across the line of regard are in error and will be discussed further in connection with visual acuity. The particular side of the target to which the oscillation is biased predominantly and the frequency of reversal of this bias varies amongst subjects and are affected by gaze angle and psychophysiological factors which commonly alter CN.

Jerk (J) nystagmus is initiated by a slow drift of the fovea off the target, followed by a fast phase (saccade) which both stops the drift and fully or partially corrects the eye position error (Dell'Osso et al., 1974). Thus, both P and J nystagmus are the result of slow eye movements off the target and both reflect an instability in the slow eye movement (SEM) subsystem (Dell'Osso & Daroff, 1974).

The genesis of CN is related to the attempt to fixate or direct the eyes and is unrelated to ambient or retinal illumination (i.e., eye lid position) (Dell'Osso, et al., 1974). Reports relating CN to either of these latter variables are misleading in that they failed to consider the key variable of fixation attempt. The confusion resulting from the contradictory findings of these observations (e.g., 'nystagmus disappeared behind closed lids but increased in the dark') can be cleared up with the realization that the observations were related entirely to fixation attempt and unrelated to the imposed conditions.

Waveform Classification

Classification includes three main groups: pendular, jerk, and dual. The jerk group is further divided into two sub-categories: unidirectional and bidirectional.

Pendular Waveforms

Fig. 1 illustrates the pure pendular (P) form of CN and contains two shifts in waveform bias from one side of the target to the other. In this and in all subsequent figures an upward deflection indicates an eye movement to the right and a downward deflection is to the left. For the first six cycles, the subject oscillates to the left of the target. This is followed by one cycle to the right, a short interval on target, and two cycles to the left. The purity of sinusoidal form is verified by the velocity waveform which, since it is obtained by electronic differentiation of the position signal, is particularly sensitive to any saccadic components that may be present. The sinusoidal nature of the velocity trace insures the absence of any small saccades hidden in the position trace; the importance of the velocity waveform will become obvious when other waveforms are discussed.

A deviant form of pure pendular CN is asymmetric pendular (AP) as shown in Fig. 2. This form usually occurs as the subject looks laterally but is occasionally present in primary position. AP is sometimes mistaken clinically for a jerk nystagmus. Further discussion of this misidentification will be presented in the Discussion.

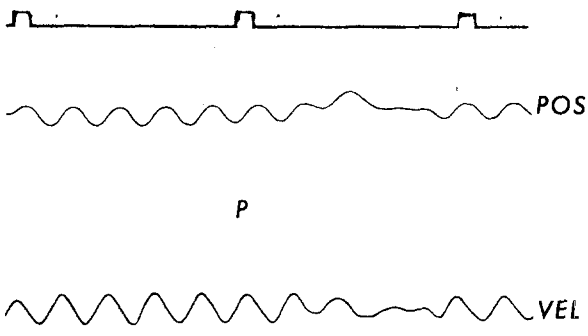


Fig. 1. Eye position (POS) and velocity (VEL) record of pendular (P) nystagmus. For the first six cycles the eyes are oscillating to the left of target (left is down and right is up). This is followed by one cycle biased to the right of target, 250 msec on target and two cycles to the left again. Note the purity of sinusoidal form in both traces and the absence of any velocity spikes. Intra- and intersubject amplitudes are variable. Timing markers are at 1 sec intervals.

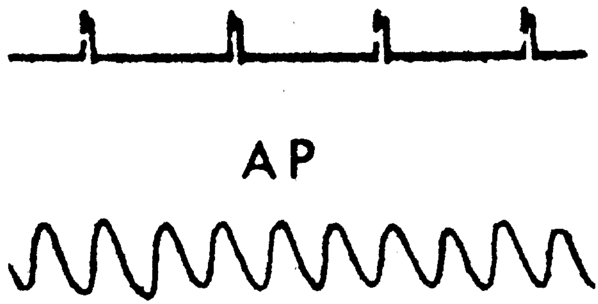


Fig. 2. Eye position record of asymmetric pendular (AP) nystagmus showing skewed sinusoidal waveform. Timing markers at 1 sec intervals.

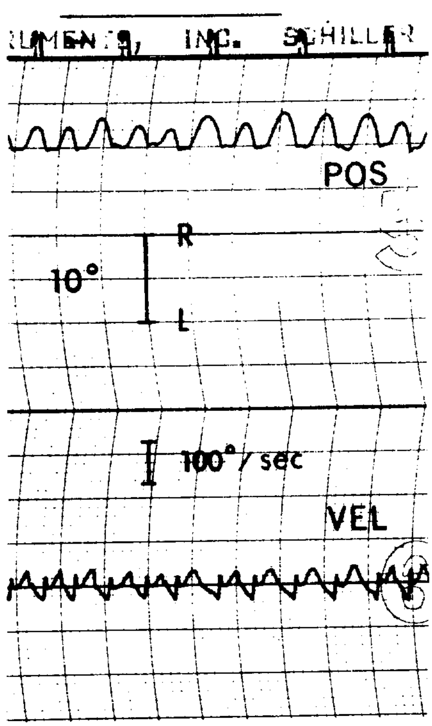


Fig. 3. Eye position (POS) and velocity (VEL) record of pendular nystagmus with foveating saccades (P_{FS}). Note flattened peaks corresponding to target foveation in this rightward-biased waveform and the velocity spikes identifying the rightward foveating saccades. Timing markers are at 1 sec intervals.

The final pendular form, shown in Fig. 3, is pendular with foveating saccades (P_{FS}). Small 'braking' saccades stop the SEM after it has bypassed the target and, in addition, the saccades achieve refoveation of the target. A flattened appearance of the waveform following the braking saccade identifies the interval of target foveation and obviously enhances visual acuity. The presence of the small foveating saccade is easily detected by the velocity tracing; the sharp rightward spike notes a saccadic eye movement. The subject oscillated to the right of the target during the entire interval.

Jerk Waveforms

As previously stated, the jerk forms of CN will be divided into two main groups based on the directionality of the saccade(s) present in each cycle. Unidirectional forms have one saccade per cycle which is always in a refoveating direction and defines the direction of the nystagmus. Bidirectional forms contain two saccades per cycle and both are in a refoveating direction.

Unidirectional jerk nystagmus can be divided into four different types. The first type is pure jerk nystagmus (J), such as that commonly seen in vestibular nystagmus. The second is jerk nystagmus with extended foveation (J_{EF}). In both, the saccades are of sufficient amplitude to fully refoveate the target. The classical saw-tooth appearance of jerk nystagmus is illustrated in Fig. 4. Identification of this jerk right (JR) nystagmus is obvious even without the accompanying velocity waveform. Fig. 5 depicts an example of J_{EF}. There are long periods of foveation following each saccade during which the eye is motionless and good vision possible. The two reversals (at the beginning and end of the figure) of the initial drift directions result in obligate reversals of the nystagmus direction as evidenced by the change in direction of the saccade spikes in the velocity tracing (two JR beats are followed by a series of jerk left (JL) and then four JR beats).

In the two other types of unidirectional jerk nystagmus, the saccades, although corrective in nature, are of insufficient amplitude to fully refoveate the target. Fig. 6 illustrates the pseudo-cycloid form (PC), in this case left pseudo-cycloid (LPC). This waveform, often misidentified clinically as pendular, consists of an accelerating rightward drift off target which is terminated by a small leftward braking saccade and followed by a SEM which refoveates the target. In some cases the small braking saccade is not clearly evident from the position record alone. However, the addition of a velocity tracing, as in Fig. 7, clearly shows the presence of even the smallest braking saccade. Note that even the two drifts off the target in a leftward direction

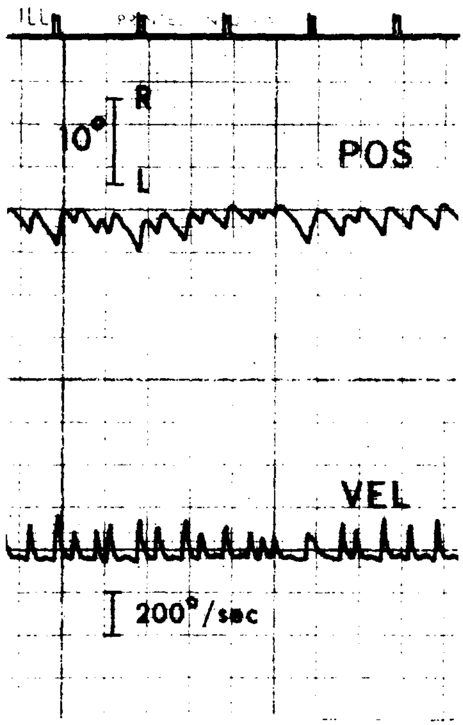


Fig. 4. Eye position (POS) and velocity (VEL) record of pure jerk nystagmus. Target foveation occurs briefly at the termination of the rightward saccade. Velocity spikes clearly identify the JR direction. Timing markers are at 1 sec intervals.

are terminated by small rightward braking saccades. Fig. 8 shows the final form of unidirectional jerk nystagmus, pseudo-jerk (PJ). In this bizarre waveform the actual direction of the 'fast' phase is opposite to the apparent direction. The names pseudo-jerk left (PJL) and pseudo-jerk right (PJR) refer to the apparent rather than actual direction. The waveform in Fig. 8 is actually a jerk left nystagmus as shown by the velocity tracing, but its clinical appearance is jerk right and it is therefore identified as PJR. This particular oscillation is initiated by a rapidly accelerating drift off the target to the right which is terminated by a small braking saccade to the left, followed by a leftward SEM that moves the eyes back on target. Because of the rapid acceleration of the initial drift off target ('slow' phase), the refoveating SEM that follows the braking saccade ('fast' phase) actually con-

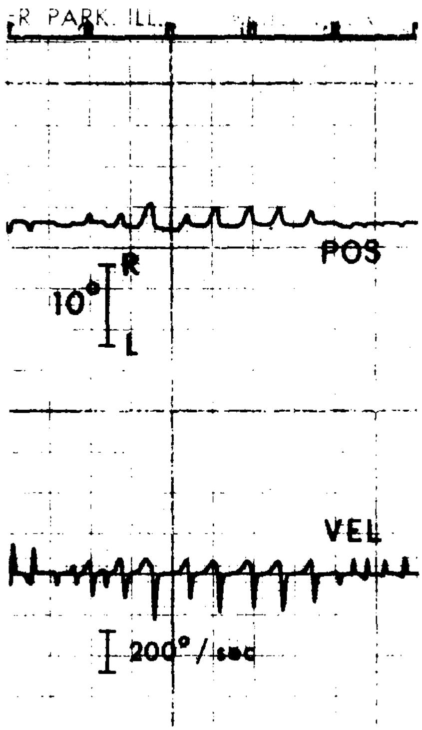


Fig. 5. Eye position (POS) and velocity (VEL) record of jerk nystagmus with extended foveation (J_{EF}). The target is foveated for the variable interval of time following the corrective saccade. Two JR_{EF} beats are followed by twelve JL_{EF} and then four JR_{EF} as is easily seen by the VEL spikes. Timing markers are at 1 sec intervals.

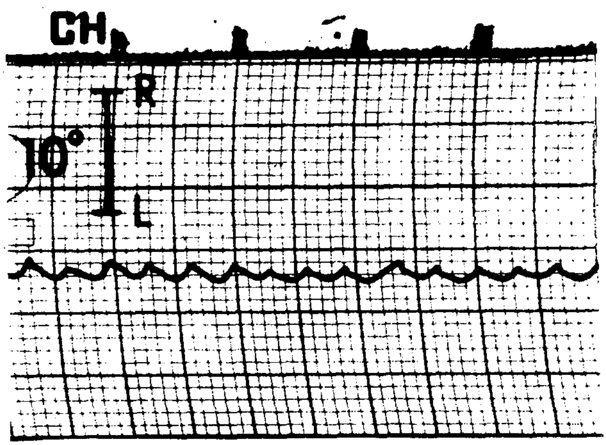


Fig. 6. Eye position record of the pseudo-cycloid (PC) form of jerk nystagmus. The leftward braking saccades in this LPC example are visible in some of the beats. Target foveation occurs at the extreme left portion of this waveform. Timing markers are at 1 sec intervals.

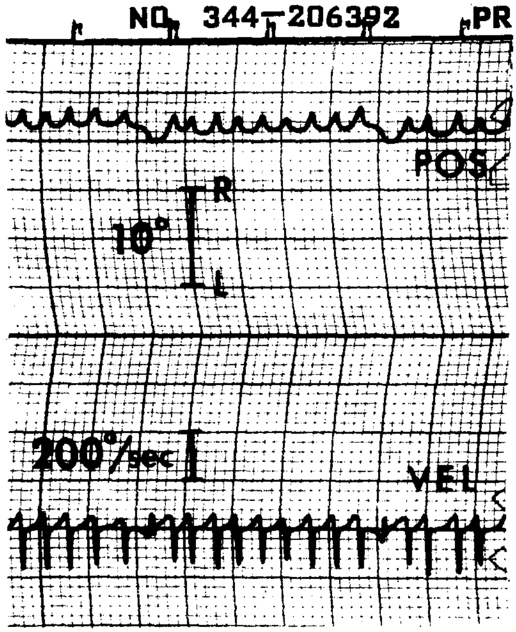


Fig. 7. Eye position (POS) and velocity (VEL) record of the pseudo-cycloid (PC) form of jerk nystagmus. Five LPC beats are followed by one RPC, nine LPC, one RPC and four LPC respectively as can be seen by the velocity spikes. Timing markers are at 1 sec intervals.

stitutes the longer time interval. This will always be clinically misidentified as a jerk nystagmus in the wrong direction because of this duration inversion of the 'fast' and 'slow' phases.

Bidirectional jerk waveforms are divided into three types and occur at gaze angles in the transition (neutral) zone in which the direction of jerk nystagmus is reversed. As such, they are transient, quite variable, and usually not conducive to good vision. Fig. 9 is a record showing several beats of pseudo-pendular (PP) nystagmus, so named because of its pendular clinical appearance and identification. At the extreme left of the figure the eye is on target and an accelerating rightward drift off target begins. It is terminated by a leftward braking saccade which initiates a leftward accelerating SEM. The eye passes the target and continues leftward until a right-

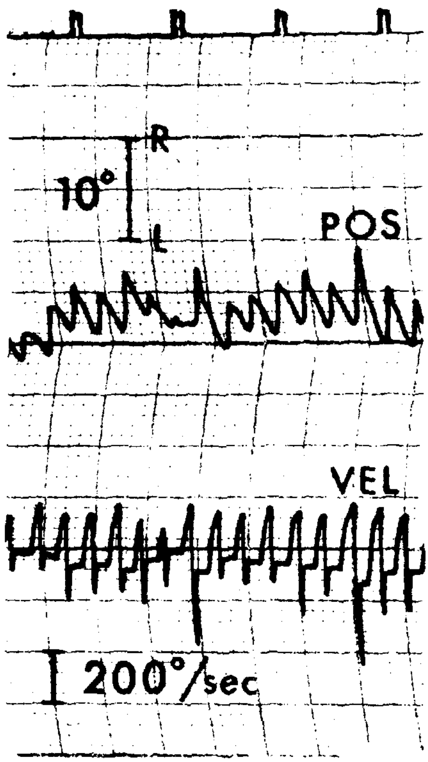


Fig. 8. Eye position (POS) and velocity (VEL) record of the pseudo-jerk (PJ) form of jerk nystagmus. These PJR beats are easily identified by the leftward VEL spikes although the waveform appears clinically to be JR. Timing markers are at 1 sec intervals.

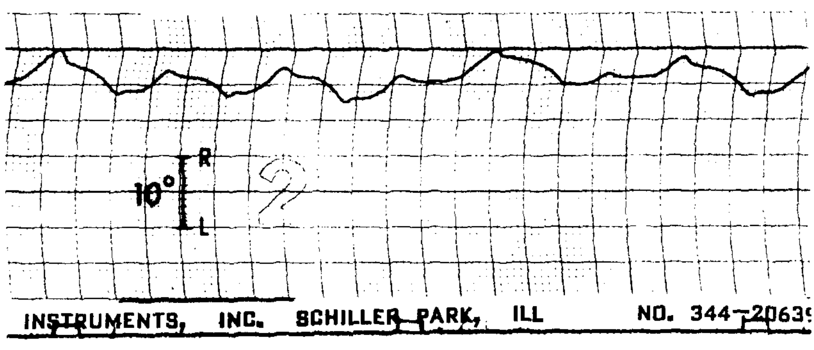


Fig. 9. Eye position record of the pseudo-pendular (PP) form of jerk nystagmus containing the characteristic bidirectional braking saccades. The beat starting at the second timing marker is biased fully to the right with target foveation occurring at the extreme left peaks. Timing markers are at 1 sec intervals.

ward braking saccade stops this SEM and initiates a rightward accelerating SEM. In this way, with alternating accelerating SEM and braking saccades, the PP waveform is generated. The target can be somewhere between the peaks of the oscillation or at one of the peaks as shown by the fifth cycle of Fig. 9. The characteristic pattern of alternating saccades is seen clearly in Fig. 10 where, after three LPC beats, four PP are shown followed by two more LPC beats.

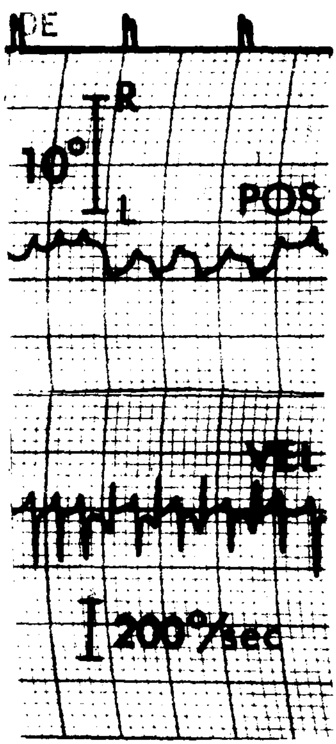


Fig. 10. Eye position (POS) and velocity (VEL) record showing four pseudo-pendular (PP) cycles preceeded by three left pseudo-cycloid (LPC) beats and followed by one LPC. The alternating VEL spikes clearly identify the PP waveform. Timing markers are at 1 sec intervals.

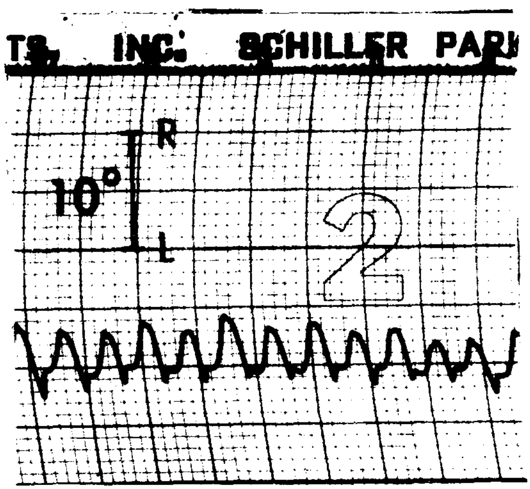


Fig. 11. Eye position record of the pseudo-pendular with foveating saccade (PP_{FS}) form of jerk nystagmus. In this example, the PP_{FS} waveform is biased to the right of target with the rightward foveating saccades and following flattened peaks identifying the interval of target foveation. The smaller leftward braking saccades at the right peaks of the waveform are visible in some of the cycles. Timing markers are at 1 sec intervals.

A sub-group of PP contains foveating saccades (PP_{FS}). Here, as shown in Fig. 11, the waveform is biased (as is P_{FS}) such that the braking saccades that actually foveate the target are larger than those at the other peaks. They are also of variable amplitudes depending upon the actual overshoot of the target. This is clearly demonstrated on Fig. 12 which includes the velocity waveform. In this tracing the subject has just executed a rightward saccade and is settling in on a new target position which is at the leftward peaks of the PP_{FS} waveform and the rightward peaks of the RPC beats which are interspersed toward the end of the tracing.

When bidirectional jerk waveforms occur at the termination of SEM that are not rapidly accelerating, a triangular waveform (T) results as can be seen in two beats of Fig. 13. The T waveforms are easily distinguished from the LPC beats surrounding them in Fig. 13 and have the same alternating spike appearance of their velocity traces as do PP.

The final bidirectional jerk form (BDJ) is demonstrated in Fig. 14. A direction is assigned to this waveform since one of the two saccades actually foveates the target. The waveform in Fig. 14 is BDJR since the eye maintains target foveation for short intervals after the rightward saccades. This is particularly evident after the large rightward refixation saccade (accompanied by an eye blink) where a BDJR is followed by two JR_{EF} and finally another BDJR waveform which terminates on the target.

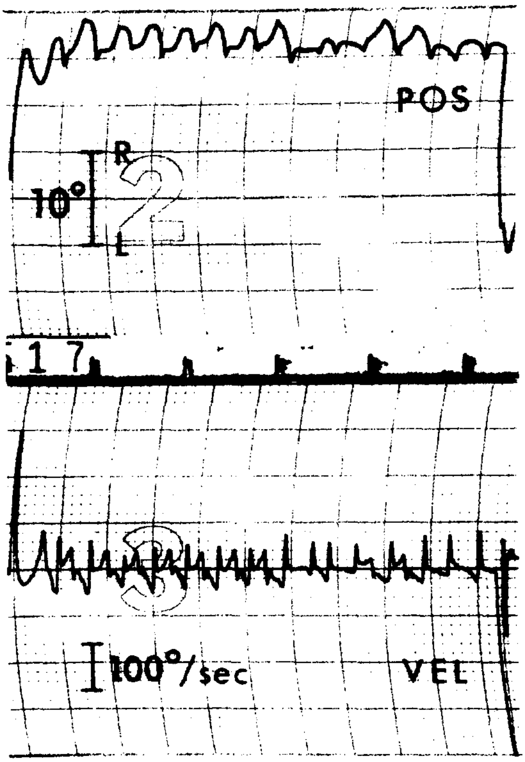


Fig. 12. Eye position (POS) and velocity (VEL) record showing a voluntary rightward saccade, eight pseudo-pendular with foveating saccades (PP_{FS}) beats, two right pseudo-cycloid (RPC) beats, two PP_{FS} cycles, three RPC beats and voluntary leftward saccade respectively. Both the larger rightward foveating saccades and smaller leftward braking saccades of the PP_{FS} waveform are easily identified by their respective VEL spikes. Timing markers are at 1 sec intervals.

Dual Waveforms

Dual jerk (DJ) nystagmus consists of the simultaneous admixture of jerk and pendular nystagmus with the superimposition of a rapid small amplitude sinusoidal oscillation upon the larger amplitude jerk nystagmus. Three cases are depicted in Fig. 15 demonstrating various degrees of the pendular oscillation, apparent on the SEM, ranging from gross (a) to moderate (b) to fine (c).

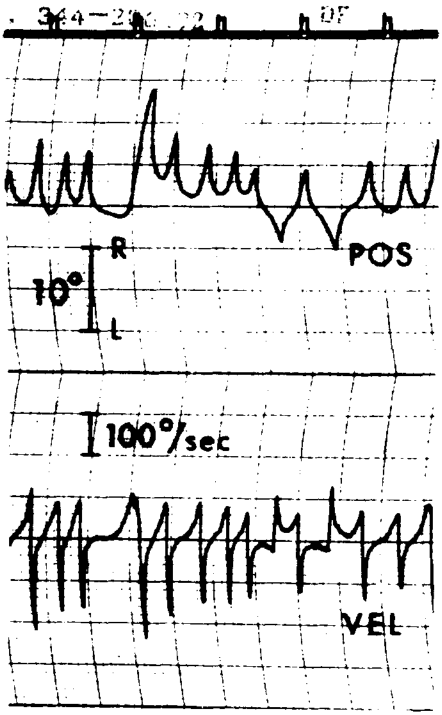


Fig. 13. Eye position (POS) and velocity (VEL) record showing two triangular (T) beats interspaced between left pseudo-cycloid (LPC) beats. The T beats have the same alternating VEL spikes characteristic of pseudo-pendular cycles. Timing markers are at 1 sec intervals.

DISCUSSION

The observations derived from the utilization of accurate eye movement recordings and laser-target retinal cinematography have made the limitations inherent in the oversimplifications of the classical descriptions and definitions of congenital nystagmus rather apparent. The use of vague definitions as guidelines for clinical observations has led to misinterpretations and erroneous identifications of waveforms. Without accurate position and velocity waveforms, certain types of CN cannot be correctly identified. The problem begins at the very definitions of pendular (P) and jerk (J) nystagmus which, based upon clinical observations alone, are invalid. For example, the notion that movements of 'equal speed' in either direction is a sufficient

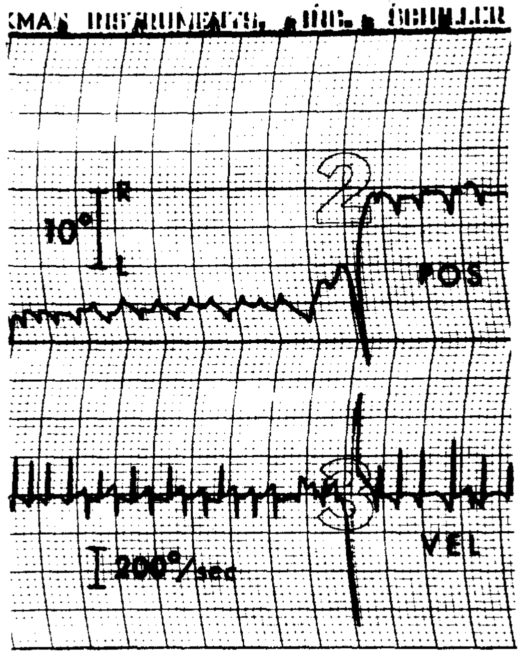


Fig. 14. Eye position (POS) and velocity (VEL) record illustrating the occurrance of bidirectional jerk (BDJ) nystagmus. This record contains four jerk right with extended foveation (JR_{EF}) beats followed by three BDJR beats, one left pseudo-cycloid (LPC) beat, one BDJR beat, three LPC beats, one BDJR beat, a voluntary rightward saccade (and simultaneous eye-blink), two JR_{EF} beats and one BDJR beat, respectively. The VEL spike patterns are invaluable in distinguishing these multiple forms of jerk nystagmus. Timing markers are at 1 sec intervals.

criterion to identify a waveform as pendular, obviously fails to deal with PC and PP types or even square wave jerks (Gegenrücke) (Jung & Kornhuber, 1964). Similarly, the use of terms such as 'fast phase' and 'slow phase' is inadequate in the cases of PC and PJ as well as the bidirectional forms PP, PP_{FS}, T and BDJ.

Based upon our waveform investigations, we propose the following two definitions for the broad categories of P and J nystagmus:

Pendular — An ocular motor instability of the SEM subsystem resulting in periodic motion of the eyes away from and back to the intended gaze angle (or target) such that the waveform is approximately sinusoidal. Occasionally small breaking saccades will be present on the peaks corresponding to target foveation.

Jerk — An ocular motor instability of the SEM subsystem resulting in a periodic drift of the eyes away from the intended gaze angle (or target) which requires a saccade in the opposite direction to stop the SEM. The saccade may either fully refoveate the target or begin a SEM in the proper direction for refoveation. *The direction of the jerk nystagmus is defined as the direction of this corrective saccade.*

The key concepts in these definitions are: both types of waveform result from the same type of ocular motor instability (i.e., SEM); both types of waveform cause the eyes to move away from and back to the target; both types may contain small braking saccades which sometimes also achieve target foveation; and the direction of jerk nystagmus is always identified by the corrective saccadic direction independent of its actual foveating ability or the length of time required for foveation. These definitions correct previous errors and are consistent with our documented data on CN waveforms. Furthermore, they permit a meaningful systematic classification of CN.

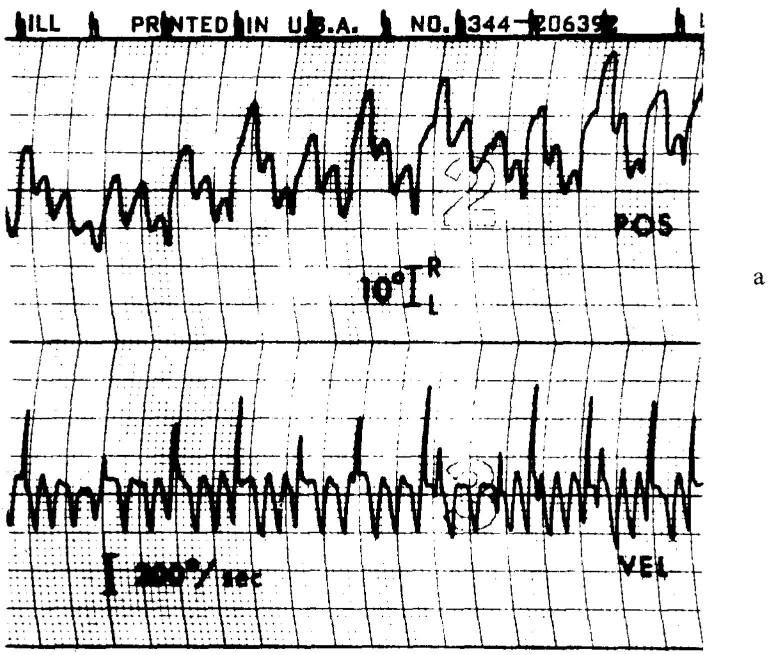
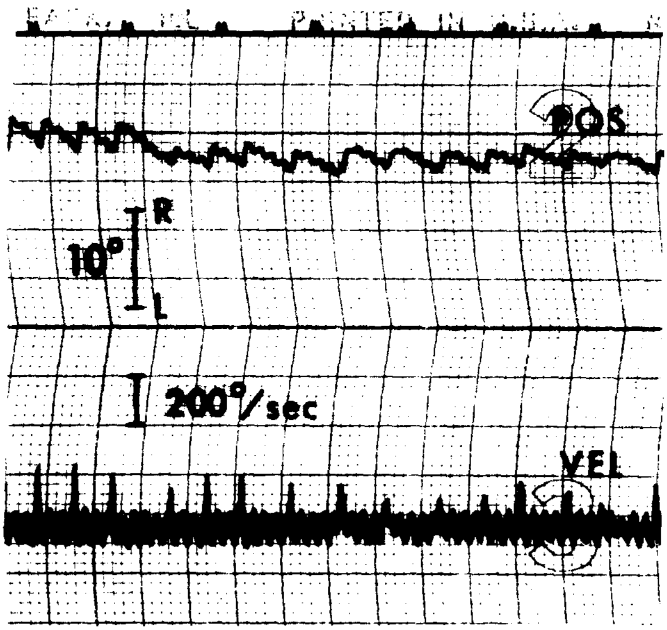


Fig. 15. Eye position (POS) and velocity (VEL) records illustrating dual jerk (DJ) nystagmus containing (a) gross (b) moderate and (c) fine pendular components. All three examples are DJR waveforms. The VEL waveforms clearly differentiate the two components. Timing markers are at 1 sec intervals.



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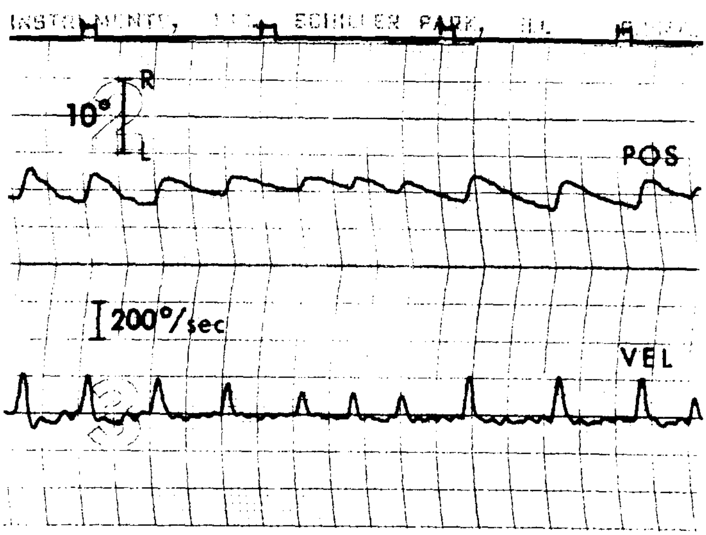

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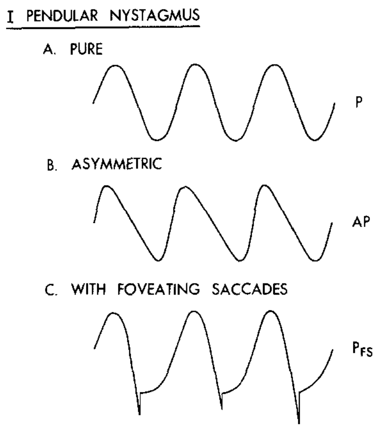

Fig. 16. Illustration of the three types of pendular nystagmus: pure (P), asymmetric (AP) and pendular with foveating saccades (P_{FS}). Note that although the foveating saccades vary in amplitude they all return the eyes to the same point (the target).

Fig. 16 illustrates diagramatically the three types of pendular CN (P, AP, and P_{FS}). Clinically, AP is sometimes misidentified as jerk nystagmus despite the absence of any saccades. Note the variability of the foveating saccades in the P_{FS} waveform. Their amplitude is dependent on the amount of overshoot of the target and all return the eyes to the target. The control system implications for this are clear. Since the actual brainstem command for each of the foveating saccades occurs prior to its execution, the required information for the calculation of its amplitude cannot be proprioceptive or visual but must be based on an internal output monitor (Weber & Daroff, 1972) which is monitoring not only eye position but also eye velocity. In this way, the amount the eyes will overshoot the target can be predicted and a saccade of the appropriate amplitude programmed to occur at the proper instant to return the eyes to the target.

Fig. 17 illustrates the four types of unidirectional jerk nystagmus (two with saccadic foveation and two with SEM foveation). Starting with pure jerk and working down to pseudo-jerk in either directional column, the key factor is seen to be the simultaneous degeneration of the corrective saccade and acceleration of the SEM off target. Thus, each type is merely a variation of the classical saw-tooth jerk waveform and contains the same saccade following the defoveating SEM instability. Specifically, in the J type, a SEM off target (usually not as linear as depicted) is followed by the direction-defining saccade which refoveated the target. In J_{EF}, target foveation is maintained for a small time interval before the SEM instability causes an

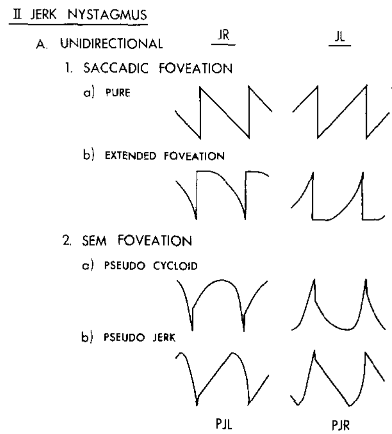

Fig. 17. Illustration of the unidirectional types of jerk nystagmus including two with saccadic foveation (pure jerk and jerk with extended foveation) and two with slow eye movement (SEM) foveation (pseudo-cycloid and pseudo-jerk). Note the reduction in and variability of saccadic amplitude in the pseudo-cycloid waveform and further reduction in the pseudo-jerk waveform.

acceleration of the eyes away from the target. This is halted and the target refoveated by the corrective saccade which defines the nystagmus direction. In the PC waveform the SEM instability causes the eyes to accelerate off target as before but the variable amplitude braking saccade which halts this is insufficient to fully refoveate the target. It is, therefore, followed by a decelerating SEM which brings the eyes back on target. The direction of this nystagmus is defined as the direction of the corrective saccade independent of its amplitude. The final waveform, PJ, starts with a rapidly accelerating SEM off target that is terminated by a small braking saccade in the corrective direction. The target is refoveated by means of a SEM of variable linearity as in the case of PC. Consistent with all unidirectional CN waveforms, the nystagmus direction is the direction of the corrective saccade and is not related to the time intervals of the two 'phases'.

The bidirectional jerk nystagmus waveforms are illustrated in Fig. 18. These waveforms are encountered in the neutral zone of gaze angles where jerk direction is in transition. With the possible exception of PP_{FS} and BDJ they generally do not support good visual acuity because of low foveation time per cycle. All four types consist of either accelerating or linear SEM which pass in alternate directions through the target. These SEM are terminated by braking saccades of alternating direction which are always corrective in nature and which may actually foveate the target (PP_{FS} and BDJ).

The braking saccades have been exaggerated in Fig. 18 for illustrative purposes. Clinically, all will be mistaken for pendular nystagmus as evidenced by the familiar reports of a 'pendular region' between the two oppositely-directed regions of jerk nystagmus in the typical subject with a diagnosis of congenital jerk nystagmus. What is actually occurring in this region of unstable equilibrium is alternate runaway of the SEM subsystem in each direction. As gaze is directed away from this neutral zone (which may be eccentric to primary position), a type of unidirectional jerk nystagmus predominates. Indeed, we have never recorded a patient with congenital jerk nystagmus who had a pure pendular waveform in such neutral zones.

Fig. 19 shows the components of dual jerk nystagmus (DJ). Basically, it consists of a variable-ampliturde, high frequency pendular oscillation superimposed on a higher amplitude jerk nystagmus. The relationship between the two instabilities presently is unclear. Convergence may damp both components simultaneously. In one patient with periodic alternating dual jerk nystagmus, the pendular component remained constant during the jerk component's cessation as the transition from one jerk direction to the other occurred.

On the basis of our classification of congenital nystagmus and identification of the possible clinical misinterpretations, the compositive CN waveforms which yield a clinical picture of 'pendular' or 'jerk' nystagmus

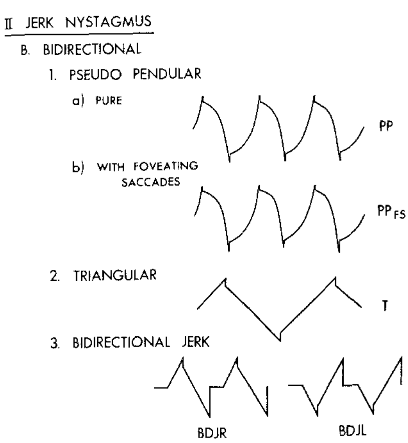

Fig. 18. Illustration of the four types of bidirectional jerk nystagmus: pseudo-pendular (PP), pseudo-pendular with foveating saccades (PP_{FS}), triangular (T) and bidirectional jerk (BDJ). All saccades are in a corrective direction (i.e., towards the target). The foveating saccades of PP_{FS} vary in amplitude but all achieve target foveation.

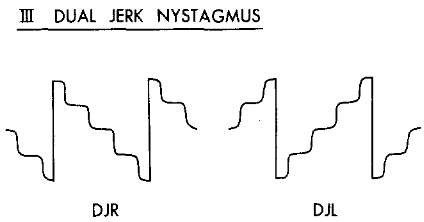

Fig. 19. Illustration of dual jerk nystagmus showing sinusoidal modulation of the slow eye movement off target.

emerges. Fig. 20 and 21 illustrate the range of waveform types with associated gaze angles which would categorize a subject (for simplicity, neutral zones are shown at O°). The clinical tendency to identify as pendular a case with a seemingly large neutral zone (in which one or several of the bidirectional jerk waveforms actually predominates) is particularly common, especially if the subject happens to have an associated visual defect. For this reason, it becomes exceedingly difficult to evaluate studies and reports which claim causality between certain visual defects and nystagmus waveforms determined entirely by clinical observation. There is no assurance that any or all of those subjects identified as having pendular CN actually had one of the P waveforms. Indeed, previous investigators who have utilized eye movement recording have failed to find any causal relationship between primary visual disturbance and CN (Jung & Kornhuber, 1964).

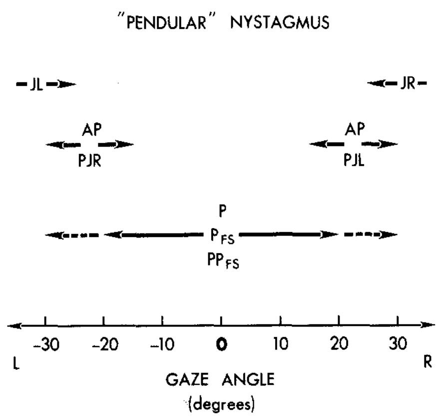

Fig. 20. Illustration of the ranges of various nystagmus waveforms which contribute to the clinical identification of pendular nystagmus. In addition to the pendular waveforms (P, P_{FS}, AP) is the jerk waveform PP_{FS} which, if it occupies a broad range, is used to misidentify as pendular a waveform which becomes PJ and/or J on lateral gaze. The regions are not sharply defined and do overlap.

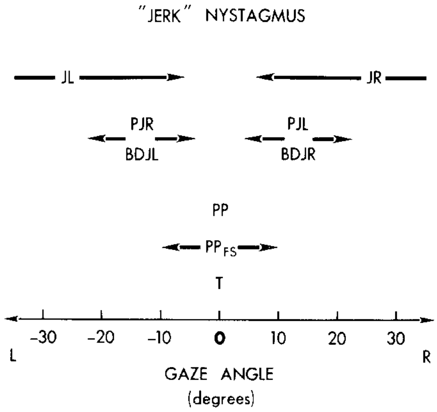

Fig. 21. Illustration of the ranges of various nystagmus waveforms which contribute to the clinical identification of jerk nystagmus. Narrow neutral zones containing the PP, PP_{FS} or T waveforms and their transition to PJ and/or BDJ and/or J on lateral gaze are the key to this clinical picture.

In the Appendix which follows, the classification of CN waveforms will appear in Table A1. We will therein also discuss waveforms chosen to illustrate the intra-subject variabilities and the foveation strategies which result in frequent bias reversals and waveform distortions. The waveforms depicted in Fig. 1–15 were chosen as examples of typical analogues to correspond to the pure waveforms illustrated in Fig. 16–19. They do not contain some of the more bizarre shapes commonly seen when transitions from one form to another occur. These will be included in the Appendix.

The waveforms discussed in this paper exclude congenital latent nystagmus which will be presented in a future paper. We have evidence that congenital latent nystagmus is actually a position-holding instability which distinguishes it from the manifest forms of CN discussed in this report.

CONGENITAL NYSTAGMUS

WAVEFORM TYPE	SYMBOL	CLINICAL APPEARANCE
I. Pendular	P	
A. Pure	P	P
B. Asymmetrical	AP	P/J
C. With Foveating Saccades	P_{FS}	P
II. Jerk	J	
A. Unidirectional		
1. Saccadic Foveation		
a. Pure	JR & JL	J
b. Extended Foveation	JR_{EF} & JL_{EF}	J
2. SEM Foveation		
a. Pseudo Cycloid	RPC & LPC	J/P
b. Pseudo Jerk	PJL & PJR	JL & JR
B. Bidirectional		
1. Pseudo Pendular		
a. Pure	PP	P
b. With Foveating Saccades	PP_{FS}	P
2. Triangular	T	P
3. Bidirectional Jerk	BDJR & BDJL	P/J
III. Dual Jerk	DJR & DJL	JR & JL

Table A1. Congenital nystagmus waveform classification, symbols and clinical appearances.

APPENDIX

Table A1 lists the CN waveform classifications, the waveform symbols used in this paper, and the clinical appearance of each. The P waveforms are usually correctly identified by observation but some of the jerk forms are usually mistaken clinically for pendular (i.e., PC and all of the bidirectional waveforms). Therefore, it is probable that the incidence of pendular nystagmus has been over estimated along with the consequent under estimation of jerk CN. DJ waveforms are usually identified as jerk nystagmus.

In this Appendix we will present and discuss CN waveforms which are either unusual or particularly illustrative of foveation strategies or macular function. In several cases of P nystagmus we have observed bursts of P cycles interspaced with motionless target foveation. Fig. A1 shows one of these cases of P nystagmus consisting of two bursts per second with each burst consisting of two beats at 5 Hz. The individual bursts were biased to either direction (i.e., in the Figure the first two are to the right and the last burst to the left of the intended gaze angle). This waveform was recorded from a subject with tapetoretinal degeneration and congenital night blindness. We have also observed P bursts in a patient with congenital DJ nystagmus whose bursts occurred after a voluntary saccade. Another patient exhibited sporadic uniocular P bursts at 5 Hz which lasted 2–3 seconds per burst. He also had tapetoretinal degeneration.

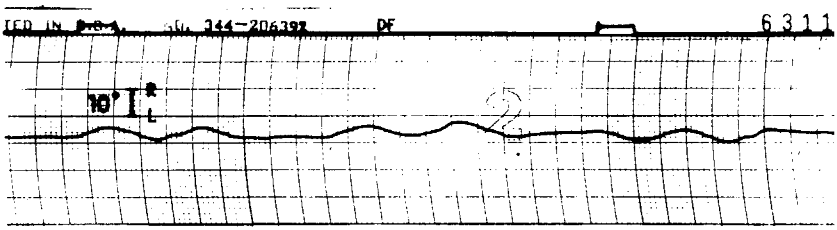

Fig. A1. Eye position record of bursts of two pendular cycles at a rate of two bursts per second. The first two bursts are biased to the right of target and the third to the left. Timing markers are at 1 sec intervals.

Fig. A2–A5 illustrate the considerable inter-subject waveform variability in jerk nystagmus. In Figure A2 there is a beat-to-beat variation between J and J_{EF} waveforms with a BDJR waveform at the third timing marker. The small leftward braking saccade at the extreme right peak of the BDJR beat is not evident in this position trace but can be seen clearly in the velocity tracing of the three BDJR beats present in Fig. A3 (timing markers 2 and 7). The small leftward velocity spikes verify the existence of the braking saccades while the large rightward spikes identify the direction of the various jerk waveforms. Such multiform variability is not unusual in jerk

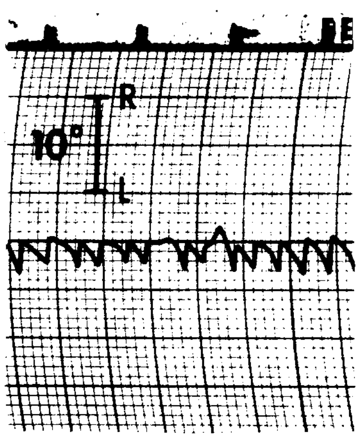

Fig. A2. Eye position record showing intrasubject variation of jerk waveforms. Beats 1, 3, 5, 7, 9, and 11–13 are jerk right; beats 2, 4, 6 and 10 are jerk right with extended foveation and beat 8 is bidirectional jerk right. Timing markers are at 1 sec intervals.

CN, an observation strongly supportive of the classification groupings we have adopted.

Fig. A4 is an expanded time scale study of the foveation strategy employed by a subject with CN and the resultant waveforms and bias reversals. To the left of the first arrow and to the right of the second arrow the nystagmus is JL_{EF}. Between the two arrows are two JR_{EF} beats. The two arrows denote the exact points where the SEM instability reverses direction. The long foveation intervals of these waveforms represents an adaptation in the developing nervous system to distort the pure form of the oscillatory instability in order to promote better visual acuity.

Fig. A5 is a typical mixture of JL_{EF} and RPC in the same subject. The sequence of beats is: $5JL_{EF}$, 1RPC, $3JL_{EF}$, 5RPC, 1 aborted JL_{EF} and 8RPC. These can be easily identified using the velocity tracing. Note the

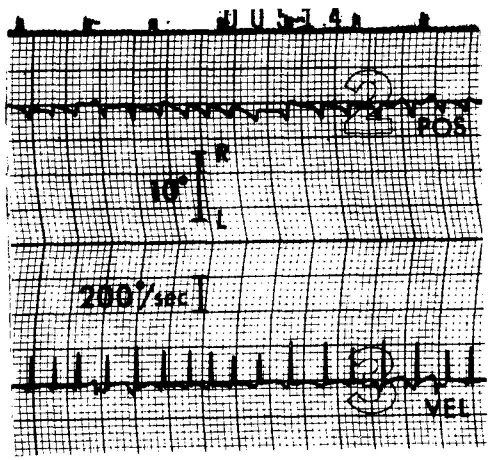

Fig. A3. Eye position (POS) and velocity (VEL) record illustrating intrasubject variation of jerk waveforms. The prominant rightward VEL spikes identify the direction of the jerk, jerk with extended foveation and bidirectional jerk (BDJ) waveforms. The three BDJ waveforms contain small leftward braking saccades identified by leftward VEL spikes. Timing markers are at 1 sec intervals.

greater velocity spikes for the larger amplitude JL saccades and also that the foveation intervals are in the same region regardless of waveform direction; such is indicative of a good foveal reflex.

The bizarre waveform of Fig. A6 consists of alternating pairs of J_{EF} as indicated in the velocity spike pairs. However, instead of a tight region of target foveation there is a band 3° wide between foveation regions of the

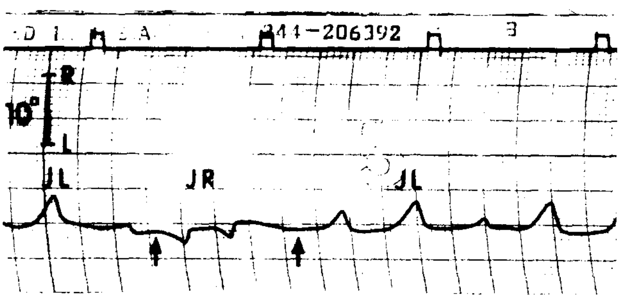

Fig. A4. Eye position record showing the slow eye movement reversals which initiate the jerk nystagmus in each direction. The reversals occur at the two arrows and the waveforms shown are jerk with extended foveation. Timing markers are at 1 sec intervals.

beats in either direction. This waveform, at first confusing, becomes understandable in light of the patient's very poorly developed fovea (as determined by ophthalmoscopic examination). Thus, instead of a sharp point of high visual acuity, his macular profile had a broad plateau of visual acuity only slightly better than the surrounding retina. Such a pattern of waveform

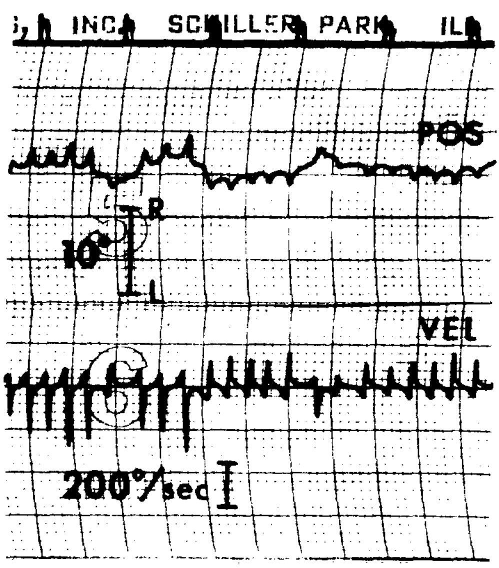

Fig. A5. Eye position (POS) and velocity (VEL) record illustrating frequent bias reversals and waveform variation in the two types shown. The leftward VEL spikes of the jerk left with extended foveation beats are larger than the rightward VEL spikes of the right pseudo-cycloid beats. Timing markers are at 1 sec intervals.

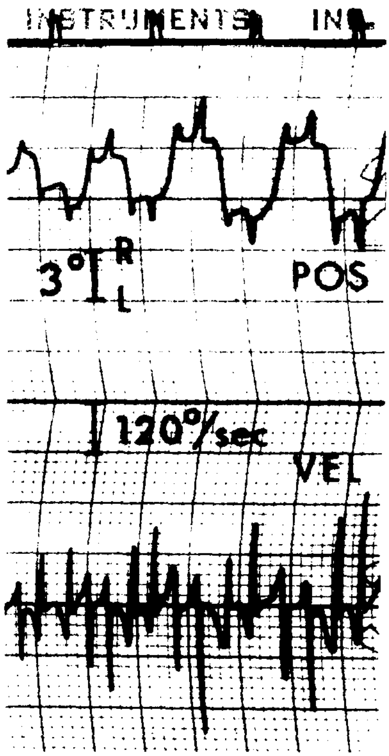

Fig. A6. Eye position (POS) and velocity (VEL) record illustrating a foveal 'dead zone' of about 3°. The waveform consists of alternate pairs of jerk with extended foveation. Timing markers are at 1 sec intervals.

reversals is indicative of the poor physiological functioning at the fovea. The waveform of Fig. A7 is from a patient who appeared, ophthalmoscopically, to have foveal aplasia. The patient had congenital periodic alternating nystagmus and it is evident from the small region of target foveation during the reversal of jerk beating directions that, physiologically, he had a functional fovea. With the use of prisms his visual acuity had improved to 20/50. Further study is required to confirm the utility of noting the bias reversals on accurate CN records and using them as an indicator of foveal function.

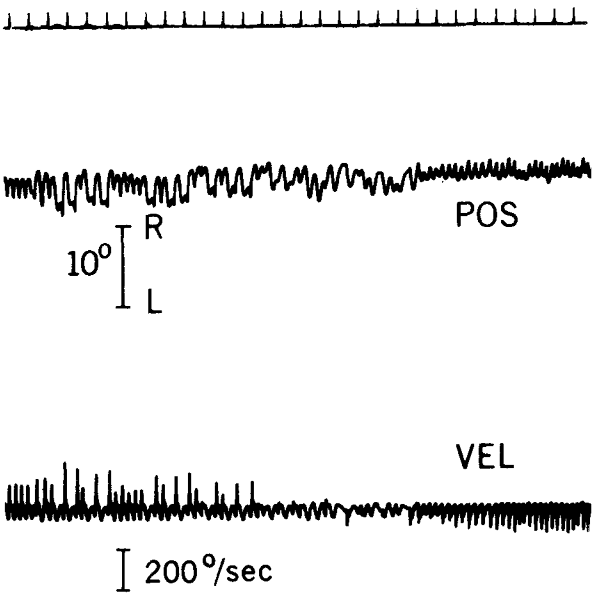

Fig. A7. Eye position (POS) and velocity (VEL) record of the direction reversal in a subject with periodic alternating nystagmus. The waveforms from both directions foveate the same target with no 'dead zone'. Timing markers are at 1 sec intervals.

ACKNOWLEDGEMENT

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Key Words

Congenital nystagmus
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nystagmus classification and definitions
eye-movement recordings and foveation strategy.