THE OCULAR MOTOR SYSTEM: NORMAL AND CLINICAL STUDIES

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In order to give an appropriate overview of our clinical studies in the ocular motor system and to exemplify the close interaction of the clinician and the bioengineer, we will provide a review of the work of the Miami ocular motor neurophysiology laboratory, including studies on subjects and patients and the implications they hold for the control of eye movements.

The framework we will present lends itself to the understanding of clinical eye signs and conforms to current research findings in this area. We have aimed at a compromise between the complexities inherent in these findings and the overly simplified presentations of most neuro-ophthalmology textbooks. The ocular motor system consists of two subsystems: version and vergence. The subsystems give rise to three types of eye movements: fast eye movements (FEM) and slow eye movements (SEM) from the version subsystem, and vergence eye movements (VEM) from the vergence subsystem. FEM and SEM are conjugate whereas VEM are disconjugate. While there are only these three ocular motor outputs, determined by spatio-temporal characteristics as well as ocular motorneuronal firing patterns, there is a multitude of input and pathological stimuli which may elicit them.

The stimulus usually employed in the laboratory to elicit FEM is target displacement. The particular FEM which results is called a voluntary saccade (Dell'Osso and Daroff, 1974). The latency is approximately 200 milliseconds, and velocity varies from 30-700 degrees/second and duration from 30 to 100 or more milliseconds dependent on the amplitude of the saccade, which normally varies between 0.5 and 40 degrees of visual angle. The

movement is conjugate and ballistic, and the control system is discrete. By that, we mean that even though there is continuous visual information, motor commands are made at some point in time and are by and large irrevocable. The control signal is retinal error. Fig. 1 is a template upon which the various pathways are superimposed, showing FEM as a closed-loop system. With head velocity 0, relative eye position in the head is the same as absolute eye position, and target displacement gives rise to retinal error. A conjugate retinal error sensed by the cortex causes signals to be sent down to the Paramedian Pontine Reticular Formation (PPRF) to initiate the eye movement and reduce the error to 0, in a simple negative feedback system.

Slow eye movements are typically generated in the laboratory during pursuit of a moving target. The resulting SEM is called smooth pursuit (Dell'Osso and Daroff, 1974). Thus, the stimulus is the target motion and the latency between onset of target motion and the movement itself is about 125 milliseconds. The velocity is usually less than 50 degrees/ second. The movement is conjugate and smooth and the control system in this case is continuous, rather than discrete. The control signal is retinal error velocity, sometimes called slip velocity. Referring back to our template (Fig. 1) we have the same interaction of eye position and target position, but a retinal error velocity signal is generated and transmitted to the cortex. A conjugate retinal error velocity again results in signals to the PPRF where a pursuit movement (SEM) is generated.

Another type of SEM (i.e., not smooth pursuit) results from vestibulo-ocular input. The stimulus is head or body motion. The latency is 100 milliseconds or less and velocity may achieve 400 degrees/second. The movement is conjugate and smooth, control is continuous, and is triggered by head acceleration. Fig. 2 is a block diagram which shows the vestibular input summing with the SEM input and FEM velocity commands, and depicts a hypothetical final common integrator. Such an integrator is required for each of these systems, as is the path around the integrator to the ocular motor nuclei and muscle plant. The fast and slow systems are within a feedback loop, but the vestibular system is not. On our template (Fig. 1) head acceleration is shown to cause vestibular input to the brainstem; the resultant output is absolute eye position which is the sum of relative eye position and head position. It is open loop; even though we've indicated the mathematical relationship between head acceleration and head position, this is not a physiological path. If vision is also present, the visual feedback path is closed aroung this loop and the resultant is a combination of open-loop and closed-loop systems operating synergistically.

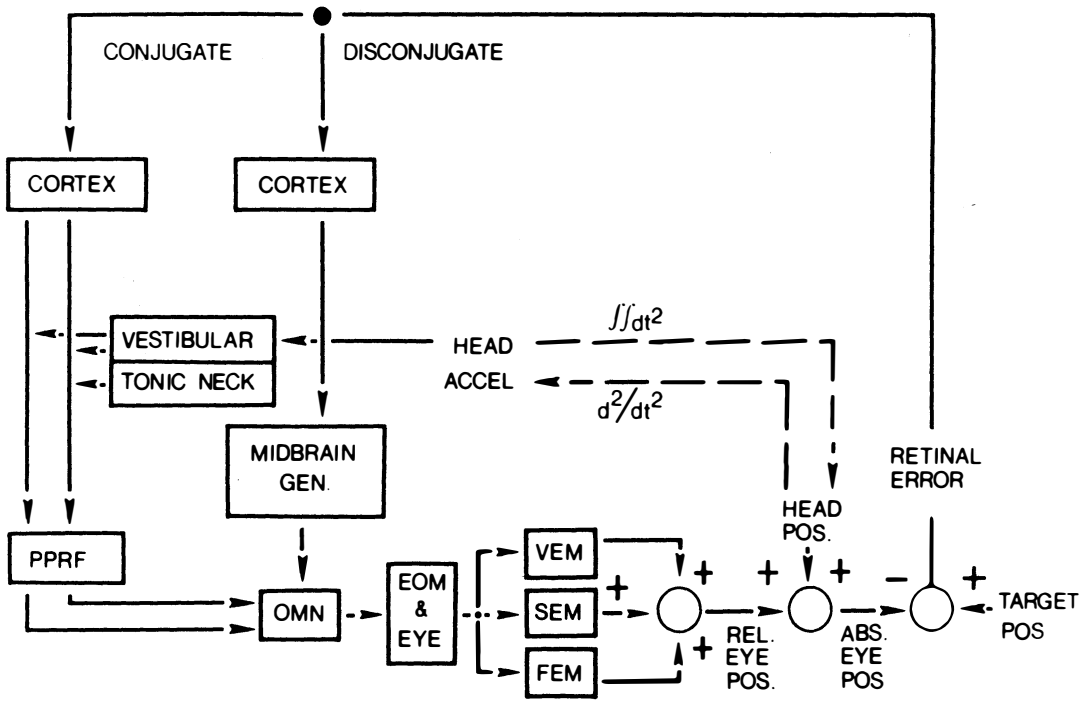

FIGURE 1. Basic block diagram of the ocular motor system with vergence and dual-mode version subsystems. Explanation of the various components are provided in the text.

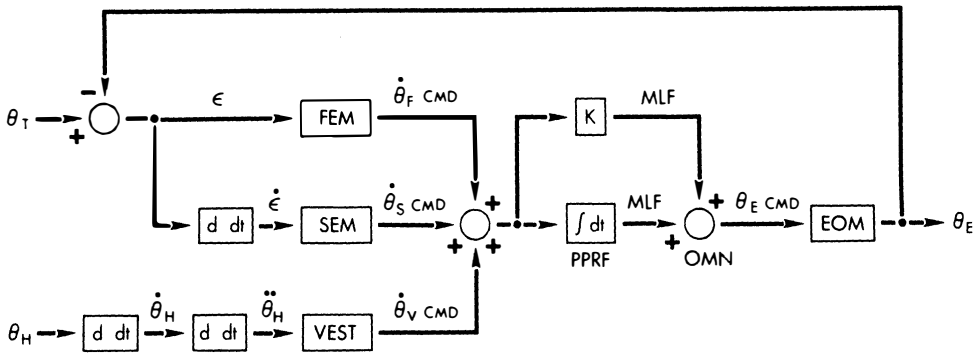

FIGURE 2. Block diagram of the dual-mode version subsystem with vestibular input which illustrates the difference between the closed-loop fast eye movement (FEM) and slow eye movement (SEM) mechanisms and the open-loop vestibulo-ocular apparatus. For simplicity the velocity commands of the FEM ($\dot{\Theta}_F$ CMD), SEM ($\dot{\Theta}_S$ CMD), and vestibular eye movements ($\dot{\Theta}_V$ CMD) are shown summing and utilizing a final common integrator ($\int$ dt) located in the PPRF. Its output and the velocity outputs travel to the oculomotor nuclei (OMN) via the medial longitudinal fasciculus (MLF). The eye position command (Θ_E CMD) is sent to the extra-ocular muscles (EOM) to effect the required eye position (Θ_E). Θ_T is the target position. In this way, the position error, $\varepsilon=\Theta_T-\Theta_E$ and the velocity error, $\dot{\varepsilon}=\frac{d}{dt}(\Theta_T-\Theta_E)$, are driven to zero; there is no feedback to the vestibular system, which responds to head acceleration ($\ddot{\Theta}_H$). Head position (Θ_H) and velocity ($\dot{\Theta}_H$) are also shown along with their relationship to $\ddot{\Theta}_H$.

The stimuli for vergence eye movements are target displacement or target motion in the Z direction, that is, toward or away from the subject. The latency is 160 milliseconds and the velocity is usually less than 20 degrees/second. The movement is disconjugate but smooth, the control is continuous, and the control signals are either retinal blur or diplopia, or both. Retinal blur is an open-loop input and diplopia is a closed-loop input. On the template (Fig. 1) is the representation of diplopia. A disconjugate retinal error is sensed and gives rise, in the midbrain, to commands to move the eyes disconjugately and thereby reduce the diplopia to 0 by the same negative feedback. In the following material we will look at various clinical abnormalities, and try to relate them to malfunction of the ocular motility control systems.

Referring to Fig. 3, disorders of the FEM subsystem results in signs like opsoclonus, dysmetria, Gegenrücken, macro square wave jerks, and macro saccadic oscillation. Nystagmus (either congenital or acquired) is due to problems of the SEM system, and vestibular malfunction will give rise to vestibular nystagmus. Internuclear ophthalmoplegia (INO) has been discussed previously in this symposium and the implicated lesions in the medial longitudinal fasciculus (MLF) are indicated in Fig. 3.

The earliest studies in our laboratory defined the metric characteristics of horizontal saccadic eye movements in normal adult humans (Weber and Daroff, 1971). Recording from each eye simultaneously, an EOG analysis of horizontal saccades defined their metric characteristics and detailed the trajectory relationships during refixation. Nine distinct left eye - right eye combinations are recognized and analyzed. (Table 1.)

Eye movements that were accurate were defined as normometric, those with errors, as dysmetric. An inverse relationship between amplitude and accuracy emerged as the basic principle of saccadic metrics. Ten degree saccades were normometric in the majority of trials; the remainder primarily represented conjugate under/or overshoot. However, the frequency of normometric saccades decreased significantly at 20 and 30 degrees. As the amplitude increased, conjugate undershooting became more prevalent. Of paramount importance was the finding of disconjugate or disjunctive eye movements, such as involved in a left eye undershoot with simultaneous right eye overshoot. Such observations demonstrate variability and frequently only approximate equality in yoke muscle performance during horizontal saccades. The most common difference in the performance of the two eyes resulted from a monocular error.

Analysis of these errors revealed a distinct tendency toward adductor overshoot or abductor undershoot. Disconjugate eye movements occurred in 15% of all trials, although the percentage in individual subjects varied from 2.0 to 26.2 per cent.

All the dysmetric saccades were followed by a small corrective movement which accomplished alignment of the fovea with the new fixation target. Analysis of these correction movements for both conjugate and disconjugate refixational errors was reported by Weber and Daroff (1972).

Two types of corrective movements (CM) occurred. One designated saccadic CM, was fast, had a definite latency, and always followed conjugate errors. The other was slow, drift-like, without a latency, and corrected disconjugate refixations. The term glissadic CM was given to the latter variety.

The saccadic correction was distinctive, easily recognized, of equal amplitude in both eyes and followed all conjugate errors. The latencies from the termination of the initial movement until the onset of the corrective movements were approximately 125 msec. The latency was similar to saccadic corrections for undershoots (positive CM) and overshoots (negative CM). The size of the CM increased monotonically from refixations of 10^{o} to 30^{o}. The glissadic correction was of low velocity (approximately 20 deg/sec) and inseparable from the terminal portion of the saccade. There was therefore no latency between the end of the initial saccade and the beginning of the correction. Thus two distinct varieties of CM were defined. These findings suggested certain mechanisms to explain the corrections accompanying both the conjugate, and in particular the disconjugate, error. We attempted in our initial block diagram to explain the different mechanisms that the ocular motor control system could use to correct disconjugate errors. Our initial concept of glissades assumed that a disconjugate error would cause a corrective movement, and it appeared from the records that these were constant velocity movements rather than the exponential movements that would reflect the muscle plant. We were therefore required to explain this phenomenon in the dark as well as with vision. We could eliminate the visual loop (the retinal loop) and, since there was no latency, we could also eliminate the proprioceptive loop (that would require anywhere from 50-80 milliseconds). The task was reduced to monitoring the output of the PPRF (the commands to the motor neurons), comparing this output with the desired eye movement, and when it was decided that, although the eyes hadn't moved yet, the command was in error, initiating another command. In this case, disconjugate saccades would result and the glissade would then appear as a continuation of the saccade.

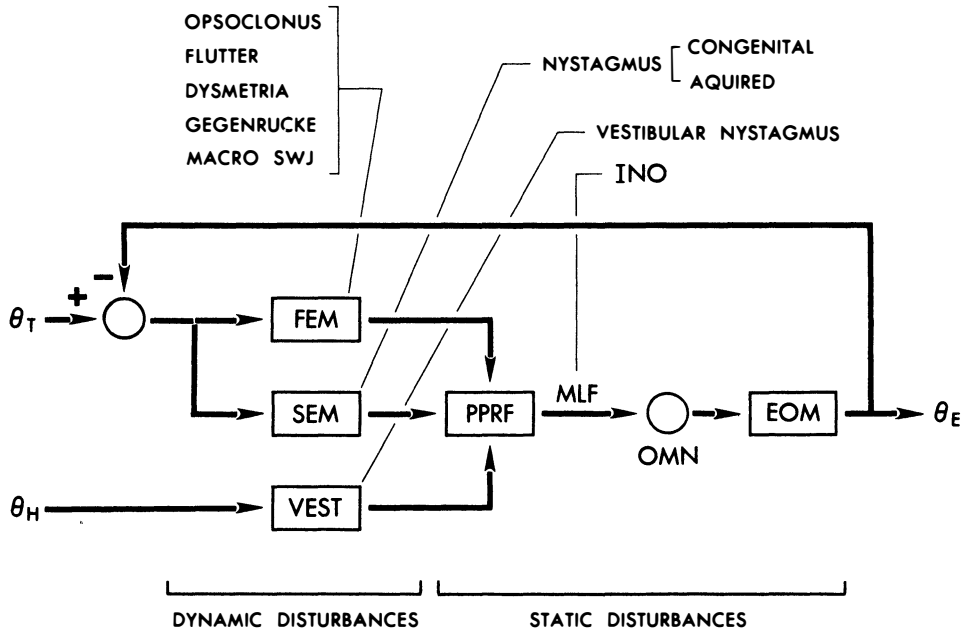

FIGURE 3. Simplified block diagram of the dual-mode version subsystem and vestibular input with various ocular motor disorders related to disturbances in specific sub-systems. Θ_T is target position, Θ_H is head position, and Θ_E is eye position.

TABLE 1. Possible Yoke-pair Combinations

(a)	LC-RC	Both eyes correct or "normometric"
(b)	LO-RO	Binocular conjugate overshoot
(c)	LU-RU	Binocular conjugate undershoot
(d)	LU-RC	Left eye undershoot
(e)	LC-RU	Right eye undershoot
(f)	LO-RC	Left eye overshoot
(g)	LC-RO	Right eye overshoot
(h)	LU-RO	Left eye under, right eye over
(i)	LO-RU	Left eye over, right eye under

As Dr. Robinson has indicated earlier in this symposium, there is some controversy as to the nature of a glissade according to its original definition by old records. It was the analysis of those records that led us to the concept of the internal monitor. A brief consideration of the anatomy underlying smooth pursuit movements will clarify the development of this concept. Earlier notions postulated either a double decussation of the pathway for smooth pursuit in the midbrain or no decussation at all. Pursuit movements are governed ipsilaterally; pursuit to the left by the left posterior hemisphere, and pursuit to the right by the right hemisphere. One of our patients had had intractable seizures as a child, and his left hemisphere had been removed 11 years previously. His refixations appeared clinically normal: however, we noted something unusual about his tracking ability (Troost, et al., 1972). Pursuit of targets moving to the patient's right was smooth, but saccadic or cogwheel pursuit occurred on following to the left. He was unable to match the velocity of targets moving in a leftward direction. We recorded eye movements during pursuit tasks and during refixations to stationary targets. We calculated the gain of his leftward pursuit and found it was low (0.2 - 0.4) despite various target velocities. While tracking sinusoids he similarly showed smooth pursuit to the right but abnormal pursuit to the left (Fig. 4).

Smooth pursuit abnormality is not of localizing value clinically when there is bilateral dysfunction (as often occurs with inattention, diffuse cerebral disease, diffuse brainstem disease or from sedative drugs). However, it is useful in pinpointing a unilateral posterior hemispheric disease to the side of the unindirectional tracking defect. There is usually a contralateral visual field defect as well.

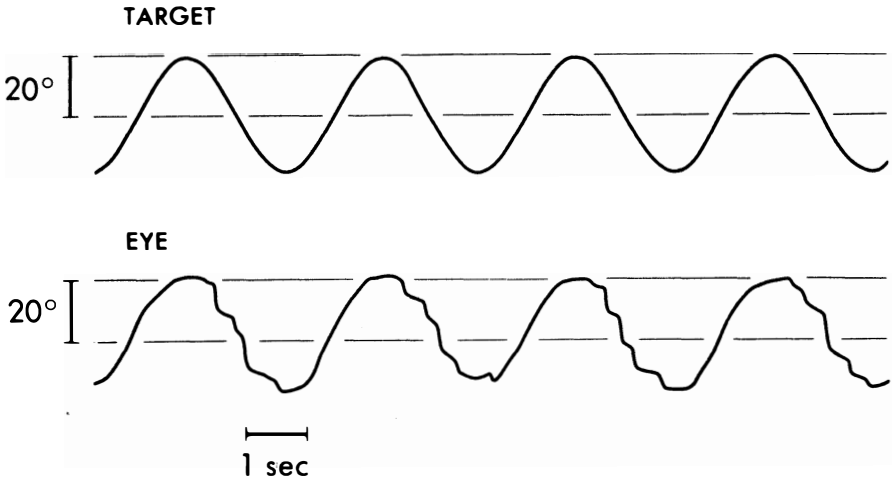

FIGURE 4. Patient with a left hemispherectomy tracking sinusoidally moving targets. Pursuit is smooth to the right (up) but saccadic or broken to the left (down).

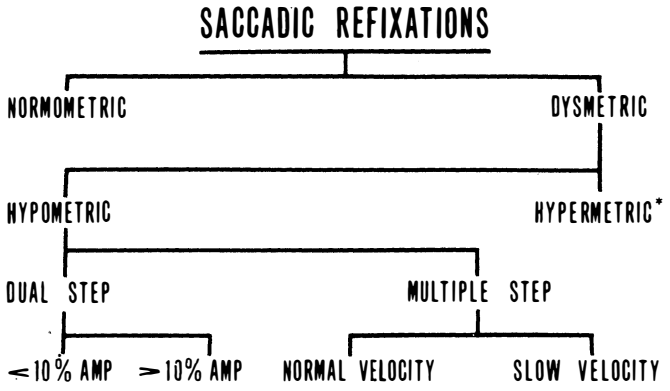

FIGURE 5. Classification of saccadic refixations. *Hypermetric saccades are unusual and have not been completely studied.

Fast eye movements were studied in the patient discussed above (Troost et al., 1972a). The pathway for FEM descends from the hemisphere and crosses in the caudal midbrain or upper pons to the contralateral PPRF. How would an individual with a missing left hemisphere make voluntary saccadic movements? Normally the right hemisphere would produce rapid eye movements in the leftward direction only; however, in this situation the right hemisphere would direct "rehabilitated" saccades in a rightward direction as well. On comparing refixation in rightward and leftward directions the patient made normal leftward saccades, but rightward saccades were abnormal in a number of ways. Accuracy was greatly reduced and only 15% of the eye movements were normometric. A high proportion of them were hypometric, composed of a series of steps, there was a larger number of disjugate errors than is seen in normal subjects, while the velocities of the rightward saccades were within normal limits.

Hypometric saccades themselves were also the subject of analysis (Troost, et al., 1974). Refixations may be classified according to their accuracy and number of corrective movements (Fig. 5). Multiple step hypometric saccades are a common type of hypometric saccade. We consider them pathologic if the velocites of the individual segments are abnormally slow. This variety of hypometric saccade is commonly encountered in patients with diffuse brainstem disease such as Progressive Supranuclear Palsy (PSP). The patient with a left hemispherectomy frequently displayed hypometric saccades during rightward refixations, but the segments had normal velocites and would therefore be considered normal individually. However, since they appeared prominently in 30% of all rightward refixations, their frequency is abnormal.

To review the findings on the hemispherectomy patient, the intact right hemisphere generated the following types of eye movements: 1. Normal rightward pursuit; 2. Defective, low gain, cogwheel pursuit to the left; 3. Normal leftward saccades; 4. Rightward saccades of normal velocity but with great inaccuracy and a high proportion of multiple step hypometric saccades.

A saccadic eye movement has a short period of acceleration, a period of high velocity, and then a short period of deceleration. Examination of activity in the agonist and antagonist muscles reveals that the tonic activity in the agonist goes from a lower level to a higher level, and the antagonist goes from a higher to a lower level. During the saccade a burst of high frequency activity occurs in the agonist which then decays to a tonic level required to hold the eye in a new position, while in the antagonist there is a complete shutoff. It is worthy of note that there is no active breaking. The antagonist does not produce a burst to stop the eye from moving. A requisite to move this highly overdamped

plant in the manner observed is a pulse-step of innervation. If only a step is applied, a 200 millisecond exponential movement results. A pulse generator has been postulated which provides a pulse of information which is then integrated in a neural integrator pool to get the step. A step is summed with the original pulse, giving the pulse-step which we see at the ocular motor neurons. Considering saccadic movements, one notes that they vary in duration with amplitude; the larger the movement, the longer it takes. Velocity also increases with amplitude. A study of normal subjects revealed that the characteristic velocity and amplitude relationship can be expressed as monotonically increasing and saturating function (Boghen, et al., 1974). We have calculated two standard deviations above and below the mean for a large sample of subjects, and our laboratory regards any eye movements that fall below the lower border as pathologically slow. There is a great deal of variation and the variation itself grows with saccadic size. The parameters of movement are statistically similar for voluntary saccades, fast phases of optokinetics, fast phases of rotational nystagmus, vestibular and also caloric-induced nystagmus, and therefore support our thesis that fast eye movements are a single homogeneous class (Sharpe et al., 1975). For saccades greater than a few degrees in amplitude there is a small but statistically significant difference between saccade velocity in light and dark, and a similar velocity dependence on structure in the visual field. Voluntary saccades made in the dark are slower than those made in the light and the same relation applies for the fast phases of rotational nystagmus. Summarizing the motor output, the PPRF sends signals ipsilaterally to the VI nucleus and contralaterally up the MLF to the III nucleus. Although this view is oversimplified, it gives the clinician a useful frame of reference when lesions appear in various locations.

Abnormal eye movements:

We are primarily concerned here with central supranuclear defects that cause either a total gaze paralysis (saccades and pursuit movements absent) or incomplete lesions causing paresis of gaze. The saccadic or smooth pursuit systems may be affected separately according to cerebral disease. Saccadic palsy with normal pursuit occurs in both congenital and acquired ocular motor apraxia and is presumably due to an abnormality which is bilateral in the frontomesencephalic projections to the brainstem. Pure pursuit palsy with entirely normal saccades due to bilateral posterior hemispheric disease is quite rare. When both fast eye movements and slow eye movements are paralyzed, it is termed a gaze palsy.

A patient with an acquired ocular motor apraxia does not have rapid eye movements and uses head movements to make refixations

during reading. When required to make refixations during head restraint, the resultant eye movements appear unusually slow. It is unclear clinically whether such eye movements are just very slow saccades, substituted vergence movements or substituted smooth pursuit movements. Vertical eye movements, including saccades, appear frequently entirely intact, at least by crude observation.

Progressive Supranuclear Palsy is a condition characterized by axial rigidity, dementia and a progressive defect in voluntary eye movements. Pathologic examination in the disorder reveals extensive neuronal loss in the basal ganglia, in the paraventricular and periaqueductal regions and in the reticular formation of the brainstem. In some patients there may be paralysis of vertical eye movements (especially in a downward direction) with some preservation of horizontal gaze. We studied a series of such patients (Troost et al., in press), with the following findings: during fixation frequent small to and fro saccades known as square wave jerks or Gegenrücke were present in all patients. The remaining horizontal eye movements were also abnormal, having low velocities and greatly prolonged durations. With head turning the eyes rotate in the opposite direction rather than following with the head as would be the case in an intact subject who was not fixating upon a target. Clinically the refixations often appeared to be hypometric as well as slow. If an observer were not aware whether a saccade or pursuit was being attempted the resultant eye movement would appear the same. The pursuit movements are cogwheel or saccadic and the saccades are hypometric with regular, slow velocity steps. Analysis of eye movement recordings distinguished the two types of output. During a pursuit task there was usually a minimal attempt at following the target. Pursuit was of low gain (0.2 - 0.4) necessitating "catch-up" saccades which were in themselves slow and of long duration; the presence of some definable pursuit made the distinction possible between a tracking attempt and a hypometric saccade. The duration of saccades was greatly prolonged. An eye movement during an attempted 15 degree refixation had a duration of up to 200 msec.

Quantitatively the vestibulo-ocular reflex in these patients was also abnormal. A normal subject can suppress the reflex when he attempts to fixate on a target which rotates with him, but the patient with PSP is unable to do this and has an apparent obligatory compensatory eye movement opposite to the head motion; he is therefore unable to maintain fixation. A normal subject can maintain fixation during head movement by making equal and opposite movements of the eyes within the head. The patient with PSP is unable to maintain fixation; he is not able to make full compensatory eye movements.

Next we will briefly consider some aspects of internuclear ophthalmoplegia (INO). During horizontal refixations the adducting eye either fails to adduct or does so slowly. The abducting eye develops nystagmus with the fast phase in the direction of gaze, with the initial abduction saccade of normal velocity. The slowness of adduction (which may at times be quite subtle) is accentuated during the OKN test. If, for example, the patient has a left INO the left eye will be slow in adduction when the gaze is directed to the right. An optokinetic stimulus passed to the patient's left necessitates repetitive fast eye movements to the right. The right eye (the abducting eye) has normal fast phases but the left (adducting eye) is slow, the difference being readily detectable during the OKN test.

In order to understand optokinetic asymmetry, it is necessary to reconsider how fast phases are generated. We require a pulse and a step of innervation summed together. If only the step occurred without the pulse, the result would be a slow eye movement with exponentially decreasing velocity, reflecting the 200 millisecond plant dynamics. On the other hand, if we had the pulse but not the step, the eye would reach its intended position, would be unable to hold and would drift backwards, resulting in nystagmus. Considering the asymmetry of the abduction and adduction saccades, and superimposing a pursuit movement generated by an optokinetic stimulus moving in the other direction, the result is a clean, brisk, optokinetic response in the abducting eye (Dell'Osso et al., 1974). The adducting eye, still in the process of going to the right (in its fast phase) when the slow phase impinges upon it with a leftward impetus, has a smaller, flat-topped, type of nystagmus. This is the classical Smith and David optokinetic asymmetry sign.

We will now examine the waveforms that accompany congenital nystagmus (CN). We define pendular congenital nystagmus as an ocular motor instability of the slow eye movement subsystem, resulting in periodic motion of the eyes away from and back to the intended gaze angle or target (if there is one), such that the waveform described by the movement is approximately sinusoidal. Occasionally, there are small foveating saccades on the peaks corresponding to target foveation. The object of all nystagmus waveforms is to maximize time-on-target and in that way maximize visual acuity. This strategy on the subject's part must be taken into account when therapy is considered. Our definition of jerk nystagmus similarly involves an ocular motor instability of the slow eye movement subsystem. The result is a periodic drift of the eyes away from the intended gaze angle or target, but a saccade is required in the opposite direction to stop the slow eye movement. We term these breaking saccades. The saccade may either fully refoveate the target, or it may begin another slow eye movement in

the proper direction for refoveation. The direction of jerk nystagmus is defined as the direction of the corrective saccades and it is the only waveform parameter that remains constant. All saccades in nystagmus waveforms are directed toward the target, and whether or not they achieve foveation is irrelevant. Both the classical saw-tooth type of jerk nystagmus and pure sinusoidal nystagmus are very rare in congenital nystagmus, but they are observed. Fig. 6 shows the three types of pendular nystagmus we have observed: pure pendular nystagmus, asymmetric pendular nystagmus (pure pendular sometimes converting to jerk nystagmus) and pendular with foveating saccades.

Regarding jerk nystagmus, there are eight types: four unidirectional and four bidirectional (Dell'Osso and Daroff, 1975). Of the unidirectional types (Fig. 7), the pure jerk nystagmus resembles vestibular nystagmus. If, after the foveating saccades, the eye remains on target for a certain amount of time before its exponentially increasing velocity drift off target, the result is a jerk nystagmus with extended foveation. This is conducive to very good visual acuity. In these two cases, the fast eye movement brings the eye onto the target. An insufficient amplitude would only stop the runaway which was taking the eyes away from the target, and another slow eye movement would return the eye back to the target again. There would follow a variable period on target with no motion (good visual acuity) and again, the runaway. Such a waveform is called pseudocycloid because it resembles a cycloid. The breaking saccades are of variable amplitude and are occasionally very small. If they are very small and the slow eye movement which follows is almost linear, pseudojerk nystagmus results. Pseudo, not because it is not a jerk nystagmus (it is a jerk nystagmus since the saccade is directed toward the target), but because, when examining a patient with this waveform, one invariably misidentifies its direction. Observation indicates that the fast phase takes longer than the slow phase and the direction is defined improperly. In the transition zone between full jerk left and a full jerk right nystagmus, there are many waveforms that are variants of bidirectional jerk nystagmus (Fig. 8). They are almost always called pendular and, without tracings, it is impossible to tell they are not pendular. Pure pseudopendular nystagmus is an alternate runaway in each direction, stopped by a breaking saccade at each end. Thus, there is a slow eye movement runaway to the right, a breaking saccade to the left, a slow eye movement runaway to the left, and a breaking saccade to the right. The target is somewhere in the middle, and very poor visual acuity accompanies this waveform. In pseudopendular nystagmus with foveating saccades, the target is at one peak after breaking saccades which are of variable amplitude; those at the other peaks are not. These breaking saccades not only stop the runaway but get the eye on target and there is usually a flat area, indicating

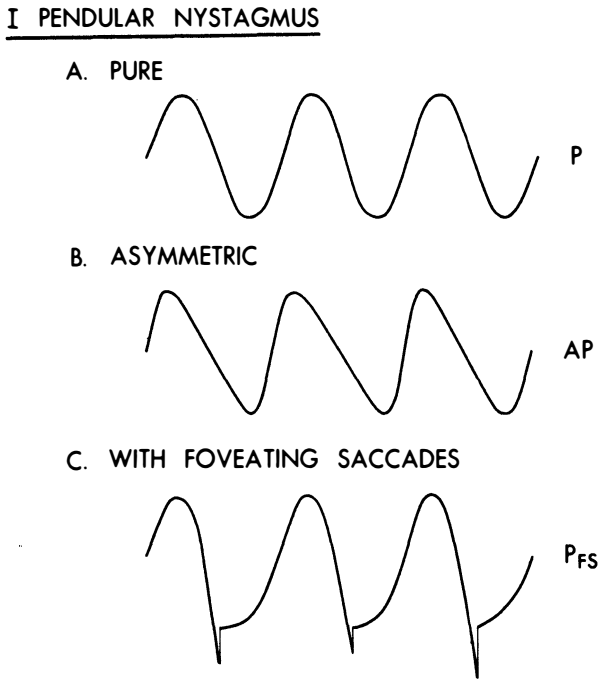

FIGURE 6. The three types of pendular nystagmus: pendular (P), asymmetric (AP), and pendular with foveating saccades (PFS). Note that although the foveating saccades vary in amplitude they all return the eyes to the same point (the target).

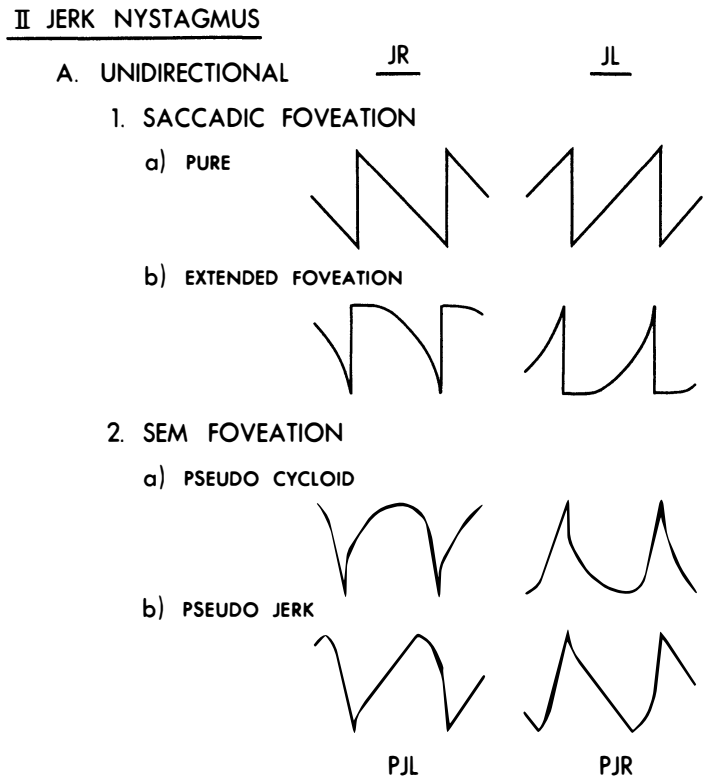

FIGURE 7. The four unidirectional types of jerk nystagmus: two with saccadic foveation (pure jerk and jerk with extended foveation) and two with slow eye movement foveation (pseudocycloid and pseudojerk). Note the reduction in the variability of saccadic amplitude in the pseudocycloid waveform and further reduction in the pueudojerk waveform.

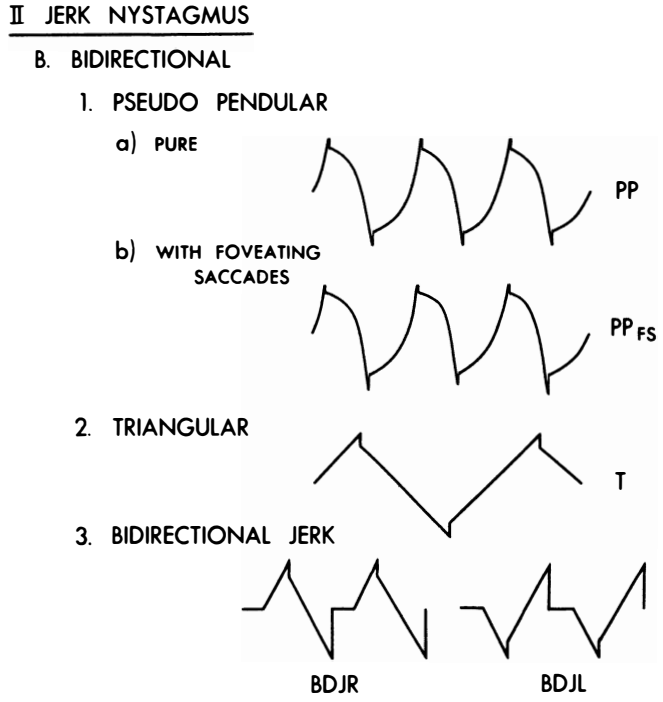

FIGURE 8. The four types of bidirectional jerk nystagmus: pseudopendular (PP), pseudopendular with foveating saccades (PPFS), triangular (T), and bidirectional jerk (BDJ). All saccades are in a corrective direction, i.e., toward the target. The foveating saccades of PPFS vary in amplitude but all achieve target foveation.

no eye movement, during which the person can see quite well. If instead of an accelerating runaway, there is a fairly linear runaway alternating in direction, we have triangular nystagmus; again with breaking saccades at both ends. Breaking saccades are always directed towards the target, which is somewhere in the middle of the waveform which is not good for vision. However, this form can be sometimes converted to one which is good for visual acuity. In such a case the saccades are large enough to achieve the target in one of the directions (assume it's the rightward direction), and there follows a long period of no eye movement with good visual acuity. Such a waveform is called bidirectional jerk right. We have given it a direction although it is bidirectional, because one of the saccades is a foveating saccade. Another type involves both pendular and jerk movements. The jerk nystagmus with pendular oscillation superimposed is usually a coarse low frequency jerk nystagmus and a fine high frequency pendular nystagmus superimposed on the slow phase and is called dual jerk right or dual jerk left (Fig. 9). These two types of nystagmus, the jerk and the pendular, are relatively independent. Sometimes convergence will abolish the jerk nystagmus; the pendular component usually remains although it sometimes also stops. The relationship of these two components is still under investigation. Clinically, a patient with CN may manifest a large zone of apparent pendular nystagmus, which could take any one of the bidirectional jerk forms, and peripherally show an apparant jerk nystagmus which may or may not be real. If he has such a large "pendular" neutral zone, the clinician is tempted to identify a case of pendular nystagmus. If on the other hand he has a very narrow neutral zone, the diagnosis may be jerk nystagmus; he may have jerk in lateral gaze and have one of the pseudopendular waveforms, which will be diagnosed as pendular, in the middle gaze positions. In the absence of eye movement recordings, the tendency therefore is to over-diagnose pendular nystagmus and under-diagnose jerk nystagmus. Consequently many speculative correlations between sensory defects and nystagmus must be re-evaluated in the light of objective criteria in the form of accurate eye movement recordings. On this basis, we have concluded that all nystagmus is a motor defect, no matter what the waveform is.

Macro square wave jerks are exemplified by a woman with demyelinating disease who has bilateral INO (Dell'Osso et al., 1975). On examination of the eye movement recordings (Fig. 10), we see that there is spurious saccade to the left, followed by a return saccade to the right after a very short latency (80 milli-seconds). The subject remains on target for a period of time before another saccade is initiated. These are macro square wave jerks; the fact that they are not very square is because the INO is bilateral. It is evident that the abducting saccades have higher velocity peaks than the adducting saccades, both eyes

abducting faster than they adduct. This instability belongs to the fast system, such that there is a spurious saccade away from the target (in this case off to the left), followed by a rapid corrective saccade to the right and finally a longer period of time on the target. After we discovered that the macro square wave jerks occurred in the dark, we knew it wasn't retinal information that was generating the corrective saccade. Nor could we depend on feedback from eye position because the conjugate return saccade would have to be programmed according to information based on asymmetric eye positions due to the INO. We were led to the model shown in Fig. 11. We postulated that the disturbance in the right PPRF pulse generator (the solid lines trace the activity) would generate the saccadic movement to the left in both eyes. This is sensed by an internal monitor that monitors the position command going to the ocular motor nuclei and compared it to the desired position of the eyes (based on retinal information). After the spurious saccade, an error is detected, sent across the other side to the pulse generator and, with very short latency, the corrective saccade (in the dashed lines) returns the eyes to target. This is an example of the application of simple models to the understanding of pathological eye movements.

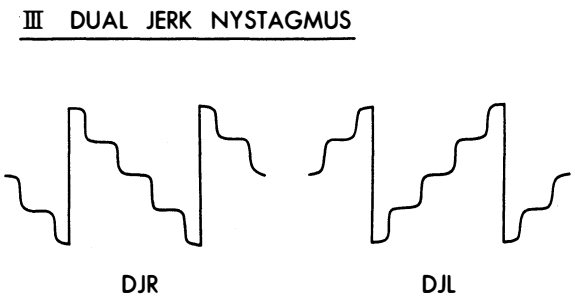

FIGURE 9. Dual jerk nystagmus showing sinusoidal modulation of the slow eye movement off target.

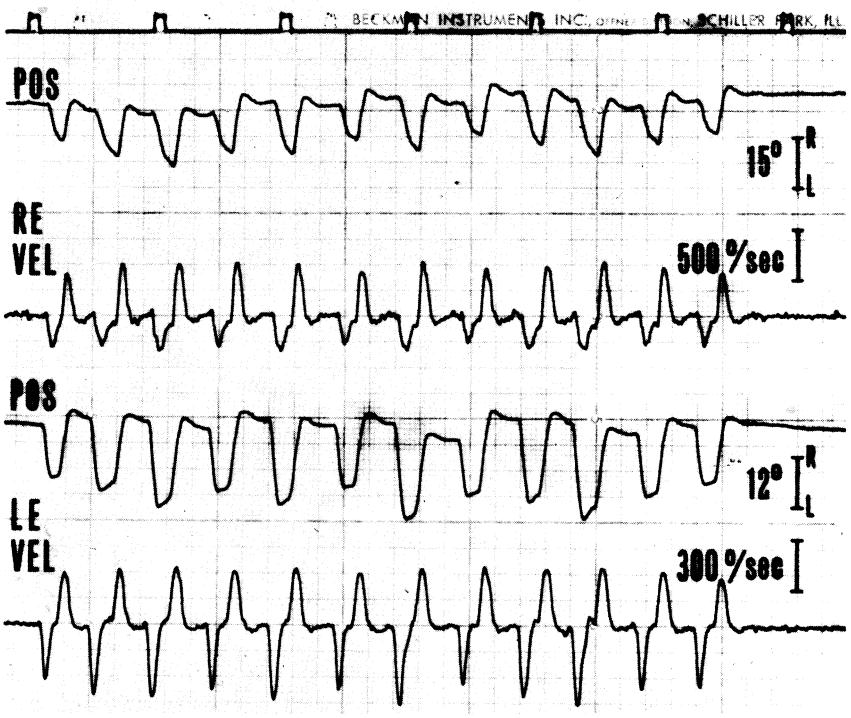

FIGURE 10. Binocular position (POS) and velocity (VEL) recordings of macro square wave jerks showing their unidirectional nature relative to the intended gaze position evident at the beginning and end of the POS traces. The oscillation consists of a leftward saccade that moves the eyes off the target and is followed, after a variable but brief latency, by a corrective rightward saccade which results in refoveation. The patient's bilateral internuclear ophthalmoparesis, with the right eye (RE) more affected than the left (LE), is apparent in both the POS and VEL waveforms. Different calibration for the two eyes should be noted and is explained in the text. The timing marks at the top indicate 1 second intervals.

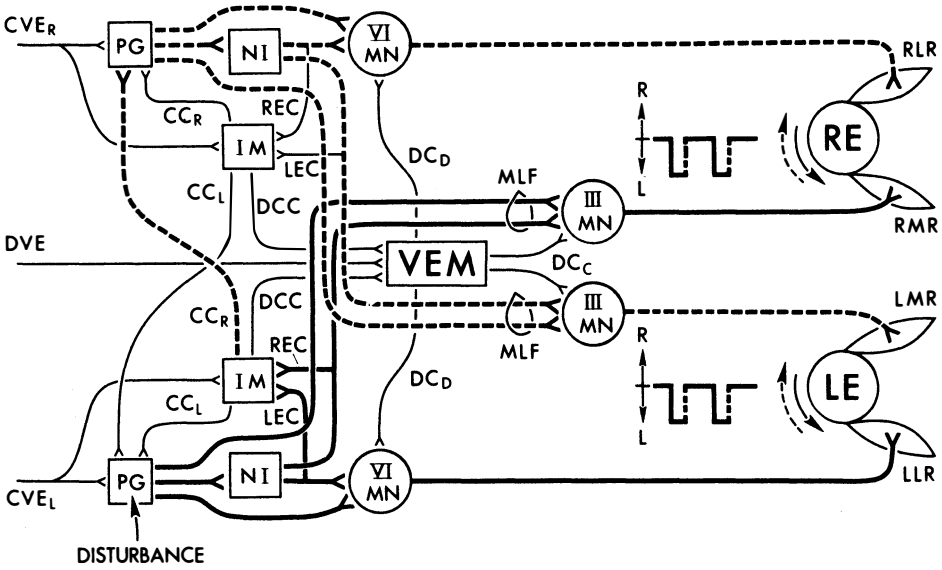

FIGURE 11. Binocular model of brain-stem output portions of the horizontal fast eye movement and vergence eye movement (VEM) subsystems illustrating the functional operation of an internal monitor (IM) in the generation of corrective eye movements. Conjugate visual errors to the right and left (CVER and CVEL, respectively) drive the pulse generators (PG) on their respective side to produce saccades.

The output of the pulse generator is integrated in the neural integrator (NI) and the resulting step of innervation is summed with the original pulse from the pulse generator at the motoneuron (MN). (Motoneuronal summing is provided for simplicity only; summing may actually occur at a prenuclear level.) Signals then go to the respective extraocular muscles (RLR, RMR, LMR, LLR) to drive the right (RE) and the left (LE) eyes. Disconjugate visual errors (DVE) drive the vergence eye movement subsystem to produce disconjugate commands of convergence (DCC) and divergence (DCD). The IM monitors the commands to both eyes (REC and LEC), compares them with the desired output (CVE), and directs the required conjugate correction to the right (CCR) or left (CCL) pulse generator as well as any required disconjugate corrective command (DCC) to the vergence eye movement subsystem. The disturbance input for this patient and the pathways for the consequent abnormal leftward saccade are in heavy solid lines, with the pathways for the corrective rightward saccade in dashed lines. The resulting macro square wave jerks are shown next to each eye. For simplicity, we have not diagrammed the internuclear ophthalmoparesis.

REFERENCES

Boghen, D., Troost, B.T., Daroff, R.B., Dell'Osso, L.F. and Birkett, J.E. Velocity characteristics of normal human saccades. *Invest. Ophthal.*, 13:619-623, 1974.

Dell'Osso, L.F., and Daroff, R.B. Congenital nystagmus waveforms and foveation strategy. *Doc. Ophthal.*, 39:155-182, 1975.

Dell'Osso, L.F., and Daroff, R.B. Functional organization of the ocular motor system. *Aerospace Med.*, 45:873-875, 1974.

Dell'Osso, L.F., Robinson, D.A., and Daroff, R.B. Optokinetic asymmetry and internuclear ophthalmoplegia. *Arch. Neurol.*, 31:138-139, 1974.

Dell'Osso, L.F., Troost, B.T., and Daroff, R.B. Macro square wave jerks. *Neurol.*, 25:975-979, 1975.

Sharpe, J.E., Troost, B.T., Dell'Osso, L.F., and Daroff, R.B. Comparative velocities of different types of fast eye movements in man. *Invest. Ophthal.*, 14:689-692, 1975.

Troost, B.T., Daroff, R.B., and Dell'Osso, L.F. Quantitative analysis of the ocular motor deficit in progressive supranuclear palsy (PSP). In press, *Trans. Am. Med. Assoc.*

Troost, B.T., Daroff, R.B., Weber, R.B., and Dell'Osso, L.F. Hemispheric control of eye movements. II. Quantitative analysis of smooth pursuit in a hemispherectomy patient. *Arch. Neurol.*, 27:449-452, 1972.

Troost, B.T., Weber, R.B., and Daroff, R.B. Hemispheric control of eye movement. I. Quantitative analysis of refixation saccades in a hemispherectomy patient. *Arch. Neurol.*, 27: 441-448, 1972a.

Troost, B.T., Weber, R.B., and Daroff, R.B. Hypometric saccades. *Amer. J. Ophthal.*, 78:1002-1005, 1974.

Weber, R.B., and Daroff, R.B. Corrective movements following refixation saccades: type and control system analysis. *Vision Res.*, 12:467-475, 1972.

Weber, R.B., and Daroff, R.B. The metrics of horizontal saccadic eye movements in normal humans. *Vision Res.*, 11:921-928, 1971.

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