ORIGINAL ARTICLE

Functional Definitions and Classification of Congenital Nystagmus Waveforms

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Although there exists abundant documentation of congenital nystagmus waveform variations in the literature, there exists no functionally relevant classification scheme of congenital nystagmus. Unfortunately, the nystagmus literature is replete with errors, misconceptions, and contradictions. Careful study of over 65 subjects with congenital nystagmus using accurate DC-coupled eye movement recording techniques and laser-target retinal cinematography has resulted in the development of composite prisms to improve visual acuity in congenital nystagmus and provide both insight into the genesis of this ocular motor instability and a sounder basis for waveform classification.[1-3] These data also have dictated tight, new, quantitatively derived definitions of pendular and jerk waveforms which are free from unreliable clinical impressions that have formed the basis for many of the above-mentioned misconceptions.

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Measurement of Velocities and Time Intervals

Paramount in the area of misleading clinical impressions are the concepts of "movements of equal speed in each direction" as an indicator of pendular nystagmus and "a fast phase in one direction and slow phase in the other" as an indicator of jerk nystagmus. First, clinical observation yields only mean velocities and time intervals rather than peak velocities or partial time intervals for different type movements in the same direction. Furthermore, these concepts are a result of an extreme oversimplification of congenital nystagmus waveforms into classical sinusoidal (pendular) or saw-tooth (jerk) waveforms. While such waveforms do occur in congenital nystagmus, they are rare.

There exist, as we will show in this paper, pendular waveforms whose directional components are of unequal speed in each direction and jerk waveforms whose directional components are of equal speed. Thus, "equal speed" cannot be used to determine waveforms; indeed, Gegenrücke (square wave jerks) are of equal speed, as are normal small bi-directional fixation saccades. Certainly we would not call these movements pendular nystagmus. Unfortunately, in a recent paper, square wave jerks have been misidentified as "saccadic nystagmus" adding more confusion to the literature.[4] Similarly, there exist pendular waveforms with directional components which appear as fast and slow phases and jerk waveforms where the time intervals of the two phases are equal; there is also a jerk waveform where the movement in the jerk direction, i.e., "fast phase," takes more time than that in the other direction, i.e., "slow phase." Needless to say, such waveforms are always misdiagnosed clinically when observation alone is relied upon and some may even be missed without eye velocity information to supplement the eye position tracing.

Given the high incidence of confusion of pendular and jerk waveforms in congenital nystagmus, and of the direction of the jerk waveform itself, all claims currently in the literature regarding etiologic inferences based on waveform, or directional effects of superimposed latent components, are highly suspect. Specifically, waveform cannot be used to infer etiology, i.e., "sensory defect" or "motor defect."

The major conclusions of our previous work will be summarized and compared with current erroneous ophthalmologic dogma.

Pendular and Jerk Nystagmus

Pendular nystagmus is a sinusoidal-like movement of the eyes biased such that the fovea rests on the target at one or the other peak of the oscillation.[2] Thus, statements depicting pendular congenital nystagmus as a to-and-fro movement across the line of regard are in error and would obviously preclude the good visual acuity possible in patients with congenital nystagmus. Both the predominant side to which the oscillation is biased and the frequency of bias reversals are idiosyncratic and subject to gaze angle and psychophysiologic factors which commonly alter congenital nystagmus.

Jerk nystagmus is caused by a slow, but usually accelerating, drift of the fovea off target, followed by a saccade which both stops this drift and either fully or partially corrects the error in the eye position.[3] Thus, both pendular and jerk nystagmus are due to slow eye movements off the target and both reflect an instability in the slow eye movement subsystem.[5]

The genesis of congenital nystagmus is related to the attempt to fixate or direct the eyes. It is unrelated to ambient or retinal illumination, i.e., eyelid position.[2,3] Previous attempts to relate congenital nystagmus to either of these latter variables were misleading in that they failed to consider the key variable of fixation attempt. The confusion resulting from the contradictory findings of such attempts (e.g., "nystagmus disappeared behind closed lids but increased in the dark") can be alleviated by the realization that the observations were related entirely to fixation attempt and were unrelated to the imposed conditions. This finding is consistent both with observations of decreased or no nystagmus in patients who are not attending to a visual input (e.g., when day-dreaming) and with the opposite condition whereby attempts to read lower on the eye chart produce intensified nystagmus and even head nodding which was absent during reading of the upper, more easily seen lines. This vicious cycle of increased effort causing increased nystagmus which decreases acuity is discussed more fully in terms of positive feedback elsewhere.[6]

New Definitions

Based upon our investigations of congenital nystagmus waveforms the following two definitions for the broad categories of pendular and jerk nystagmus are offered:

Pendular: 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) such that the waveform is approximately sinusoidal. Occasionally small foveating saccades will be present on the peaks corresponding to target foveation.

Jerk: An ocular motor instability of the slow eye movement 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 slow eye movement. The saccade may either fully refoveate the target or begin a slow eye movement in the proper direction for refoveation. *The direction of the jerk nystagmus is defined as the direction of this corrective saccade.*

The key concepts embodied in these definitions are that both congenital nystagmus types result from the same type of ocular motor instability, i.e., slow eye movement; 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 the jerk nystagmus is always identified by the corrective saccadic direction independent of its actual foveating ability or the length of time required for target foveation. These definitions correct the errors and assumptions in previous definitions and conform to documented data on congenital nystagmus waveforms. For the first time they permit a meaningful, systematic classification of this nystagmus.

Waveform Classification

Congenital nystagmus waveforms are classified into three main groups: pendular, jerk, and dual. The jerk group is further divided into two subcategories: unidirectional and bidirectional. For clarity of discussion, and in view of space considerations, each waveform type will be illustrated diagrammatically. A more comprehensive presentation including examples of each waveform and additional examples of waveform combinations is available elsewhere.[7]

Pendular waveforms: Figure 1 illustrates the three types of pendular nystagmus: pendular (P), asymmetric pendular (AP), and pendular with foveating saccades (P_{FS}). In this and all subsequent figures an upward deflection indicates a rightward eye movement. The purity of the P waveform can be verified by an eye velocity channel which would show the absence of any saccades. The AP waveform usually occurs on lateral gaze but can be present in primary position. AP is usually mistaken clinically for jerk nystagmus despite the absence of saccades. The final P waveform, P_{FS}, contains small "braking" saccades which stop the slow eye movement runaway after the eye has bypassed the target and, additionally, refoveate the target. A flattened appearance of the waveform following the braking saccade is used to identify the interval of

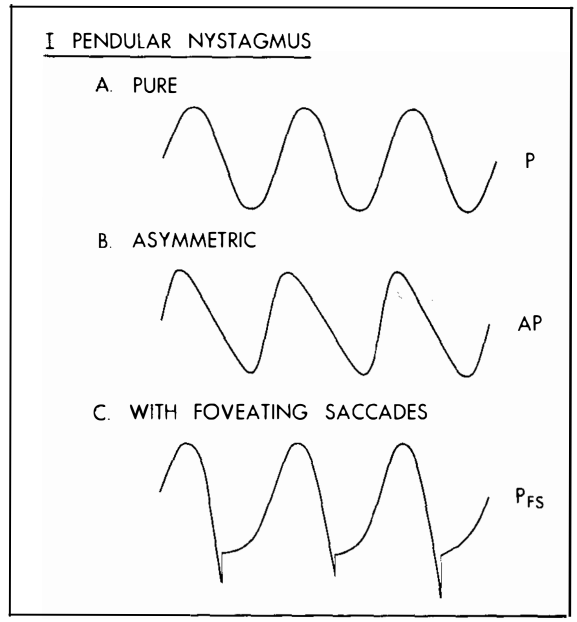

Figure 1

The three types of pendular nystagmus: pendular (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). (From Dell 'Osso and Daroff, in press.)

target foveation and good visual acuity. An eye velocity tracing easily identifies these small foveating saccades whose amplitudes vary beat-to-beat depending on the amount of slow eye movement overshoot of the target.

Jerk waveforms: As previously stated, jerk forms of congenital nystagmus are divided into two main groups based on the directionality of the saccade(s) present in each cycle. Unidirectional waveforms contain one direction-defining saccade per cycle.

Unidirectional jerk nystagmus waveforms are illustrated in Figure 2. There are four types: two with saccadic foveation (pure jerk and jerk with extended foveation) and two with slow eye movement (pseudocycloid and pseudojerk). In both pure jerk (J) and jerk with extended foveation (J_{EF}) the corrective saccade refoveates the target. In the J type, a slow eye movement off target (usually not as linear as shown) is followed by the direction-defining saccade which refoveates the target. In J_{EF}, there is an interval

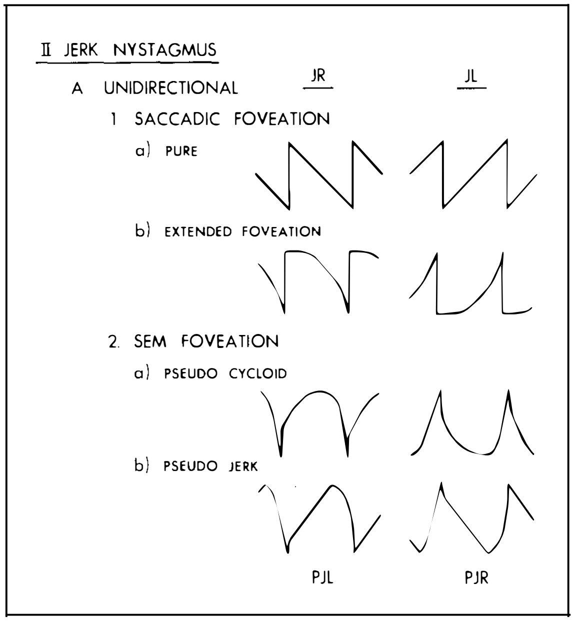

Figure 2

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 and variability of saccadic amplitude in the pseudocycloid waveform and further reduction in the pseudojerk waveform. (From Dell'Osso and Daroff, in press.)

of time during which target foveation is maintained before a slow eye movement takes the eye off target. This acceleration off target is stopped and the target refoveated by the direction-defining saccade. J_{EF} is much more conducive to good vision than J.

The saccades in the remaining two unidirectional types, although corrective in nature, are insufficient to refoveate the target. In the pseudocycloid (PC) waveform the slow eye movement instability causes an acceleration off target as before but the variable amplitude braking saccade which halts this is of insufficient amplitude for target refoveation. It is, therefore, followed by a decelerating slow eye movement which brings the eyes back on target. The direction of this nystagmus is the direction of the corrective saccade despite its small amplitude. Note that the average speed of this jerk waveform would appear clinically as equal in both directions.

The final unidirectional waveform, pseudojerk (PJ), starts with a rapidly accelerating slow eye movement off target that is terminated by a small braking saccade. A slow eye movement then refoveates the target. Consistent with all unidirectional waveforms, the direction is defined as that of the corrective saccade and is not related to the duration of the two "phases." In this case clinical impressions will always cause a misdiagnosed direction. Velocity waveforms are extremely useful in identifying the braking saccades and true direction in PC and PJ waveforms.

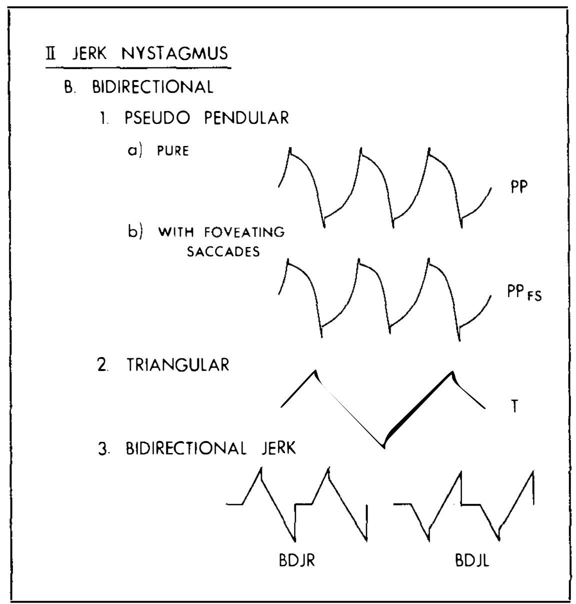

Figure 3

The four types of bidirectional jerk nystagmus: pseudopendular (PP), pseudopendular with foveating saccades (PP_{FS}), triangular (T), and bidirectional jerk (BDJ). All saccades are in a corrective direction, i.e., toward the target. The foveating saccades of PP_{FS} vary in amplitude but all achieve target foveation. (From Dell'Osso and Daroff, in press.)

Bidirectional jerk nystagmus waveforms are illustrated in Figure 3. They occur at gaze angles in the transition (neutral) zone in which jerk nystagmus reverses and, as such, they are transient, quite variable and, with the exception of PP_{FS} and BDJ, usually not conducive to good vision. All three types consist of either accelerating or linear slow eye movements which pass in alternate directions through the target. These eye movements are terminated by braking saccades of alternating direction which are always corrective in nature and which may foveate the target (PP$_{FS}$ and BDJ).

Pseudopendular (PP) consists of alternate slow eye movement runaways, each stopped by a braking saccade. The target lies somewhere between the peaks of the oscillation. A subtype of PP, pseudopendular with foveating saccades (PP$_{FS}$), is biased such that the braking saccades on one side refoveate the target. They are of variable amplitude (as in P$_{FS}$) and are larger than those at the opposite peaks.

The second type of bidirectional waveform is triangular (T). It results from fairly linear slow eye movements which pass through the target in alternate directions. Small braking saccades stop each eye movement. The final bidirectional type is bidirectional jerk (BDJ) which resembles T except that one of the braking saccades actually refoveates the target and is followed by a time interval during which the eyes are stationary and on target; this results in good visual acuity. BDJ is assigned a direction since one of the saccades actually refoveates the target.

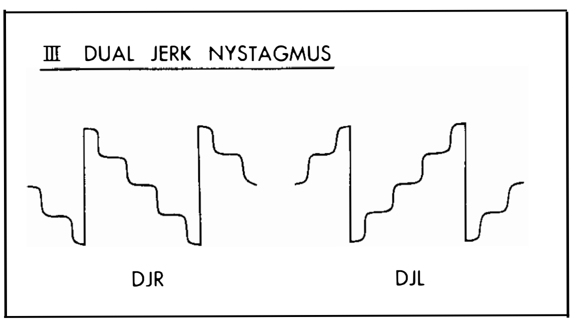

Figure 4

Dual jerk nystagmus showing sinusoidal modulation of the slow eye movement off target. (From Dell'Osso and Daroff, in press.)

Each of these bidirectional waveforms represents a region of unstable equilibrium in which there is alternate runaway of the slow eye movement subsystem in each direction. As gaze is directed away from this neutral zone (which may be eccentric), a type of unidirectional jerk nystagmus will predominate. Clinically, the bidirectional jerk waveforms will be mistaken for pendular congenital nystagmus.

Dual waveforms: Figure 4 illustrates dual jerk (DJ) nystagmus. It is a simultaneous admixture of jerk and pendular nystagmus with the assigned direction being that of the jerk nystagmus. While the pendular component may vary in amplitude, it is usually of lower amplitude and higher frequency than the jerk component. The relationship between the components is unclear. Either or both may be damped by convergence. Dual waveforms will appear clinically as jerk congenital nystagmus unless the pendular component is very large.

Summary

On the basis of these functional definitions and classification of congenital nystagmus, it becomes clear how a clinical picture of "pendular" or "jerk" nystagmus emerges. There is a tendency to identify as pendular a case with a seeming large neutral zone (in which one or several bidirectional jerk waveforms may predominate); this tendency would be irresistibly reinforced by any associated sensory defect. Similarly, the existence of a narrow neutral zone of "pendular" waveforms usually results in a clinical picture of jerk nystagmus. For this reason, it becomes exceedingly difficult to evaluate studies 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 identified as having pendular congenital nystagmus actually had one of the pendular waveforms. Studies using eye movement recordings have failed to find any such causal relationship.[8]◀

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