4.4

Nystagmus—A Contemporary Approach

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Nystagmus is a biphasic oscillation of the eyes, with the initial phase always slow (nonsaccadic), and in which the two phases have approximately equal amplitude. The word "nystagmus" is said to be derived from the Greek word *νυσταγμόσ*, meaning drowsiness which in turn is derived from *νυστάξειν*, meaning to nod in one's sleep. This derivation is curious since all forms of nystagmus are absent during sleep, a contradiction perhaps epitomizing the enigma nystagmus has represented to clinicians over the years. The venerable neuro-ophthalmologist Wilbrand[1] was quoted as advising "never write on nystagmus, it will lead you nowhere." Indeed, one of us (R. B. D.) had his early research plans discouraged by a distinguished senior neurologist who advised, "A lot of intensely intelligent and highly dedicated workers have given their lives to this subject of nystagmus, and very little has come out of it all."

With the recent surge of investigations in eye movement physiology and detailed oculographic studies of nystagmus in humans, nystagmus has become less mysterious. The necessity of quantitative oculographic studies for nystagmus research creates a heuristic dilemma for the investigator attempting to explain nystagmus to clinicians. The latter rely upon visual impressions of eye movement disorders which the investigator knows are fraught with error. None the less, rather than project nihilism and deny the utility of simple clinical observation, as opposed to oculography, we must be realistic. There are few facilities presently capable of quantitative eye movement recording and little expectation for any significant increase.

The requirements for quantitative clinical oculography are provided in Table 4.1. Alternating current systems are adequate for recording caloric-induced nystagmus but do not provide accurate eye position information and distort the wave form of linear eye movements producing a spurious exponential appearance; low bandwidth systems also distort wave forms. With EOG, bandwidths above 30 Hz lead to excessive artifact. We personally utilize an infrared system at 100 Hz. The need for the velocity information is demonstrated in Figure 4.1. Nystagmus is frequently disconjugate, necessitating measurement of each eye individually; bitemporal electrodes simply summate the two eyes and may provide totally inaccurate and misleading information. Eyelid blinks produce distortions in the horizontal recording channels that would be misinterpreted as eye movements unless detected by vertical electrodes. Rectilinear recording paper is strongly advised, as curvilinear paper makes wave form interpretation exceedingly difficult; the importance of the slow phase wave form will be explained later.

Even oculography has its pitfalls; vertical eye movements cannot be accurately measured with EOG or standard infrared systems unless the eyelids are held widely

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Table 4.1
Requirements for Quantitative Oculography

1. Direct-current EOG or infrared
2. Bandwidth of at least 30 Hz
3. Velocity channel
4. Separate measurement for each eye
5. Vertical electrodes
6. Rectilinear recording paper

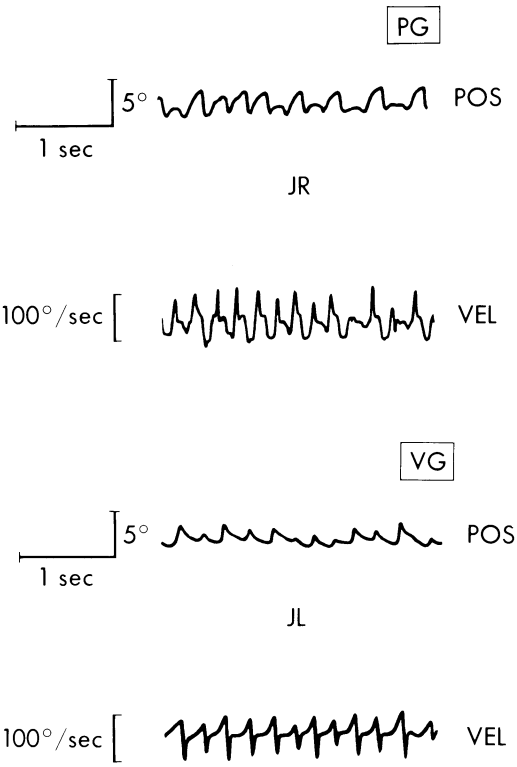

Figure 4.1. Infrared oculogram at bandwidth of 25 Hz of nystagmus position (*POS*) and velocity (*VEL*) wave forms for two patients (*PG* and *VG*). Direction of nystagmus, jerk right (*JR*) for *PG* and jerk left (*JL*) for *VG*, are identified with the aid of *VEL* tracings. The direction of the fast phases would probably have been misinterpreted with reliance entirely on the *POS* wave form. (Reprinted with permission from: L. F. Dell'Osso et al.[2]).

open mechanically. Rotary (torsional) movements are unrecordable. Thus, optimal quantitative oculography must be supplemented by careful clinical observation.

Fortunately, most forms of nystagmus and related oscillations can be recognized clinically without necessity for eye movement recording. Since many of these abnormalities have definite pathological implications, they should become familiar to the clinician.

We have recently compiled a rather complete review of nystagmus and other oscillations.[3] In this chapter we will concentrate on some specific problems. We will first present a classification, of necessity incomplete, based on mechanisms. This will be followed by a review of two important nystagmus categories, congenital and end point. We will conclude with a brief description of the clinically recognizable eye movement oscillations.

MECHANISTIC CLASSIFICATION OF NYSTAGMUS

The pathophysiological abnormalities which might explain some forms of nystagmus are high-gain instability, tone imbalance, and defective tonic innervation (Table 4.2).

1. We feel that a *high-gain instability* of the slow eye movement subsystem may be responsible for all varieties of congenital nystagmus (CN) and acquired pendular nystagmus. The evidence supporting the high-gain instability etiology of CN has been previously presented.[4, 5] Gain is defined as the output/input of a system. The most common CN wave forms are jerk, with the slow phase consisting of an increasing-velocity exponential (runaway), and pendular (sinusoidal). Control system

Table 4.2
Mechanistic Classification of Nystagmus

1. High-gain instability of slow subsystem
a. CN
b. Acquired pendular nystagmus
2. Tone imbalance
a. Vestibular nystagmus
b. Pursuit nystagmus
I. Downbeat
II. Upbeat
3. Defective tonic innervation
a. Gaze-evoked
b. Gaze-paretic

engineers have studied extensively both pendular oscillations and exponential runaways. A feedback control system is described by its system transfer function, which is the ratio of two polynomials in the variable(s) which describe input and output behavior (gain) over all frequencies. A polynomial can be factored into roots. The roots of the numerator polynomial are called "zeros," and the roots of the denominator polynomial are "poles." As the gain of a feedback control system is changed so are the positions of the poles when plotted on the s-plane (i.e., real $s = \sigma$ on the x-axis and the imaginary $s = j\omega$ on the y-axis) (Figure 4.2). The mathematical background necessary to understand clearly the position and migration of these system poles and their relationship to oscillations in a closed-loop system is too extensive to be summarized here. Figure 4.2 graphically depicts the types of oscillations which result from various pole (x) positions. Note that sinusoidal (pendular) oscillations result from $j\omega$-axis poles, and increasing-velocity runaways result from σ-axis poles. Since these poles migrate at various values of gain, it is not surprising that a system

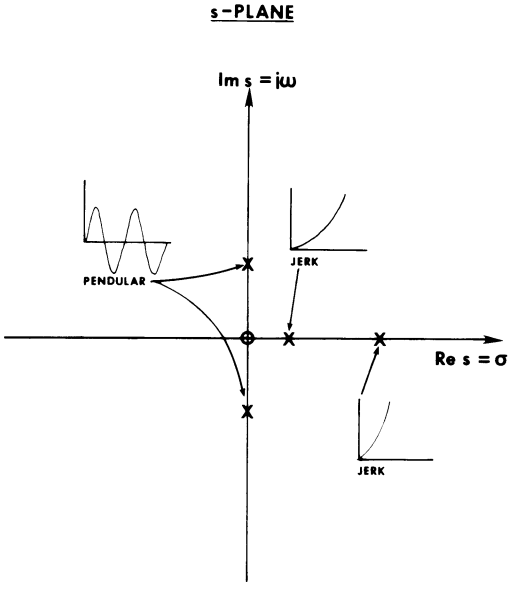

Figure 4.2. Plot of various system pole positions (×) on the complex frequency plane (*s-plane*) and the types of oscillations which result. See text for explanations.

(or a patient) may manifest both types of instability; many patients with CN have both pendular and jerk wave forms. The exact manner in which high gain affects the slow eye movement subsystem is unknown. If the instability is merely the result of high gain in a negative feedback loop, then the transfer function must be non-minimum phase (i.e., a pole or 0 must exist on the positive σ-axis). Another possible mechanism is that the rise in gain may alter the basic nature of the loop by changing a normally inhibitory junction into an excitatory one and, in so doing, create a local positive feedback loop which would allow for positive real pole positions (positive σ-axis) without having been a non-minimum phase system.

2. Tone Imbalance. In this category we include vestibular and what might be regarded as "pursuit defect" nystagmus, i.e., downbeat and upbeat. These nystagmus types are jerk and have linear (constant velocity), rather than exponential, slow phases.

a. The two vestibular end organs, with the head at rest, send approximately equal tonic spike input into the brainstem vestibular nuclei via the vestibular nerves.[6, 7] Head movement induces an inequality between the two sides which is the neural stimulus for a conjugate slow eye movement, equal in velocity but opposite in direction to the head movement; this is the vestibulo-ocular reflex. With unilateral vestibular end organ disease, there is decreased tonic spike input from the lesioned side, causing a slow ipsilateral linear drift of the eyes. Unlike the compensatory vestibulo-ocular reflex in normal situations, this ocular drift with vestibular disease moves the eyes away from the intended line of sight and is constantly corrected by repetitive saccades (the fast phases of nystagmus.) In end organ disease, the nystagmus invariably is unidirectional.

Bilateral vestibular nystagmus implies central dysfunction. The nystagmus is usually present in primary position reflecting asymmetrical involvement.

We have described the clinical phenomenology of vestibular disturbances elsewhere.[8, 9]

b. Downbeat nystagmus is defined as nystagmus in primary position with the fast phase beating downward. Zee et al.[10] demonstrated that most patients with downbeat have normal upward pursuit but are totally unable to follow smoothly in the downward direction. Instead, downward following is accomplished in a staircased fashion with repetitive saccades. The problem seemed specific for pursuit, as downward vestibulo-ocular movements were intact. These authors simulated the situation on a computer model which had normal upward pursuit velocity but abolished downward pursuit commands. The tone imbalance induced an upward drift of the eyes corrected by fast phases to return the eyes to the intended line of sight.

Upbeat nystagmus, defined as nystagmus with the fast phase beating upward in primary position, has the opposite pursuit tone imbalance (Figure 4.3). These patients can not pursue in the upward direction; the tonic imbalance produces a downward drift of the eyes corrected by the upward fast phase.

Pursuit imbalance nystagmus has not been recognized in the horizontal plane.‡ The most common cause of marked horizontal pursuit asymmetry is with posterior cerebral hemispheric disease,[11] but nystagmus is not present in these cases.

3. *Defective tonic innervation* is the cause of gaze-evoked nystagmus with a decreasing-velocity exponential slow phase. The terms "gaze-evoked" and "gaze-paretic" require clarification. Patients recovering from paralysis of gaze seem unable to maintain gaze in the direction of the previously paralysed side.[12] The eyes drift slowly toward primary position, and a corrective saccade repositions them eccentrically. Repetition of this pattern results in a gaze-evoked nystagmus aptly designated as "gaze-paretic." Gaze-evoked nystagmus is not always "paretic," which

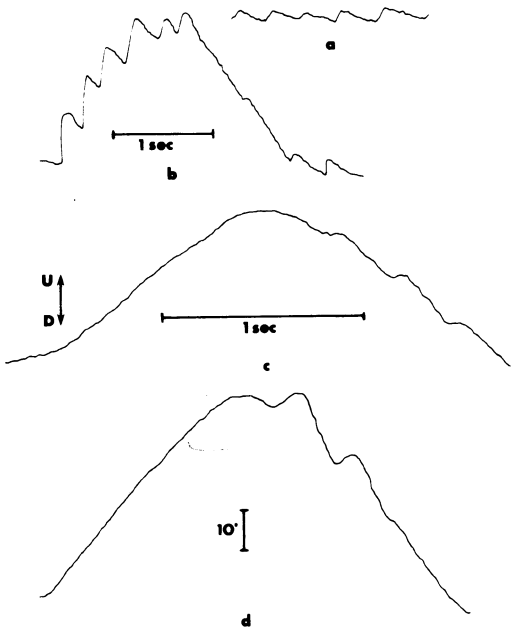

Figure 4.3. Electro-oculographic recordings in a patient with upbeat nystagmus. *a*, primary position upbeat nystagmus during straight-ahead fixation; *b*, pursuit attempt demonstrating smooth tracking in the downward direction but inability to pursue upward. The upward movement was made by a series of saccades. Between the saccades, the eyes drifted slowly downward, reflecting the omnipresent downward drift (slow phase) of the nystgmus; *c* and *d*, vertical vestibulo-ocular reflex showing presence of normal upward movement. The downward movements were too fast (gain greater than 1.0) resulting from the summation of the downward vestibulo-ocular movement with the constant downward nystagmus drift. The excessive speed necessitated the movement pauses and reversals. *U*, up; *D*, down.

prompted Jung and Kornhuber[13] to define gaze-paretic nystagmus as that type of gaze nystagmus with a low (1–2 Hz) frequency. Determination that a pulse in neural firing frequency was responsible for the initiation of saccades and that its integral, the step, was the firing pattern required for maintaining ocular deviation (see two previous chapters) inspired the obvious conclusion that an inadequate step must account for the slow phase of gaze-paretic nystagmus. The viscous damping of the orbit (plant dynamics) re-

‡ We have recently described a patient with a unilateral horizontal pursuit defect who manifested horizontal jerk nystagmus. The slow phases were linear and the fast phases bent toward the direction of the pursuit defect.[32]

quires the slow phase to be a decreasing-velocity exponential[14] (Figure 4.4), as distinct from the linear slow phase of vestibular or pursuit nystagmus. We created a relatively simple model of fast eye movements on an analog computer and simulated several inferred neuronal deficits that might be responsible for gaze-evoked nystagmus.[15]

We now suggest that the term gaze-paretic be used to describe a type of gaze-evoked nystagmus, with a decreasing-velocity slow phase.

With future advancements in our understanding of nystagmus mechanisms, the entire subject should lend itself to such classification. At present, using the mechanisms of high-gain instability, tonic tone imbalance, and defective tonic innervation, we have classified some of the major [congenital, vestibular, gaze-evoked (paretic)] and some less common (acquired pendular, downbeat, upbeat) forms of nystagmus (Table 4.2).

CONGENITAL AND OTHER NYSTAGMUS IN INFANCY

This is the area we have studied most extensively in the laboratory and our conclusions are herein summarized.

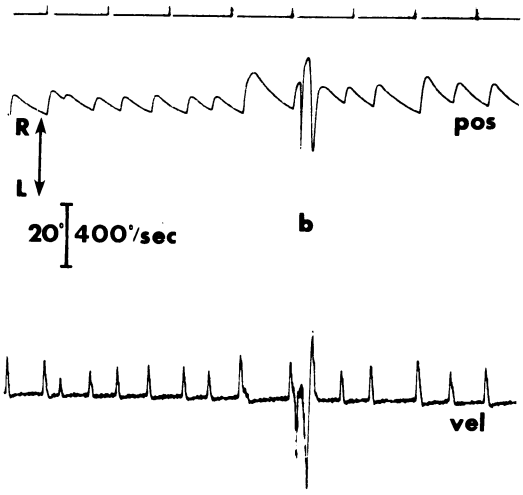

Figure 4.4. Oculographic tracing of gaze-evoked nystagmus showing decreasing-velocity exponential slow phase. *b*, blink; *R*, right; *L*, left, *pos*, eye position; *vel*, eye velocity.

1. CN, as previously mentioned, represents a high-gain instability of the slow eye movement subsystem.

2. There are many CN wave forms, and mere classification into pendular and jerk types is a gross oversimplification.[16] Moreover, the clinical separation of pendular from jerk nystagmus may be exceedingly difficult, with the bias generally being to falsely classify jerk nystagmus as pendular.

3. A primary visual abnormality in the infant or child with nystagmus should not be regarded necessarily as the cause of the nystagmus.[2] Both the visual disturbance and the nystagmus may coexist independently. The fact that visual defects are frequently present in patients with CN may represent simple genetic association. No primary visual defect can be the cause of nystagmus noted at birth, and no acquired condition (visual loss) can logically cause a *congenital* condition (nystagmus). Thus, nystagmus which does develop in a child secondary to progressive visual loss should not be designated "congenital nystagmus." The general tendency to do this has resulted in considerable nosological confusion.

4. Etiological classification cannot be based upon nystagmus wave form. Family members sharing the same genetic defect in hereditary CN may have entirely different wave form patterns.[2]

5. Nystagmus intensity is proportional to the subject's effort to see (fixation attempt); the latter is the main driving force for CN. The harder the subject tries to see, the greater the nystagmus. The increase in nystagmus further decreases visual acuity and leads to an even greater visual effort.[17] Thus, a positive feedback loop is created which reverses any willful attempt to decrease the nystagmus (Figure 4.5). Such a situation is analogous to the parkinsonian tremor, which increases when self-conciousness prompts the sufferer to attempt its reduction.

6. Many patients with CN have a distinct null region (gaze angle where nystagmus is minimum). The utilization of prism spectacles (bases left or right) to obviate the need for a head turn and reduce visual intent is of distinct benefit to these sub-

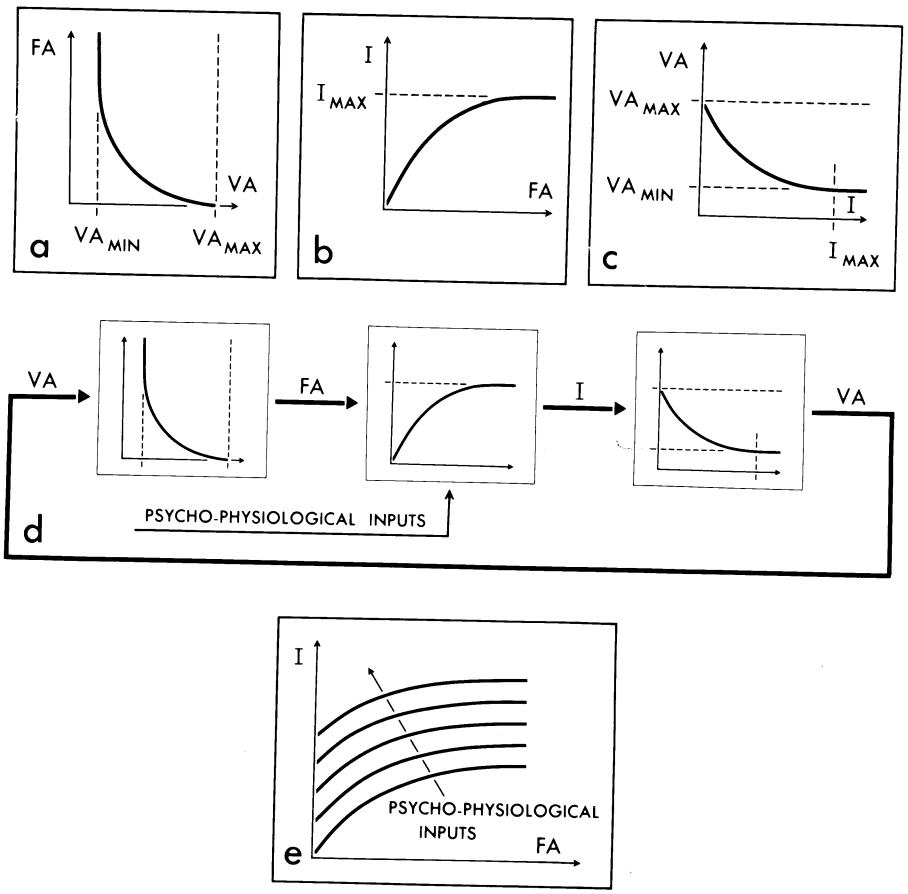

Figure 4.5. The positive feedback nature of congenital nystagmus. *a*, the relationship between fixation attempt (*FA*) and visual acuity (*VA*). With lower visual acuity, there is heightened fixation attempt; *b*, with increased FA, nystagmus intensity (*I*) increases; *c*, as *I* increases, *VA* decreases; *d*, the interrelationships of *a*, *b*, and *c* resulting in a positive feedback loop. Thus, increased *FA* lowers *VA*. *e*, the effects of psychophysiological inputs on *I* explain inter- and intrasubject variability in nystagmus intensity (Reprinted with permission from: L. F. Dell'Osso[17]).

jects. This serves to diminish the detrimental effects of the positive feedback loop depicted in Figure 4.5. Since convergence usually dampens CN, the addition of vergence prisms (bases out) to the spectacles is usually beneficial; the result is composite prisms (unequal bases out) which provide both the version and vergence shifts that combine to null the nystagmus.[4]

7. In situations where the null is too eccentric for utilization of prisms, surgical correction not only moves the null closer to primary position but also tends to flatten and extend it over a larger number of degrees, thereby minimizing the off-null nystagmus. Contrary to previous reports, we have found this result to be long-lasting.[18] Furthermore, there is increased acuity even in those cases where no preoperative increase was demonstrated at the null angle (preferred head turn).

Our present classification of nystagmus in early life is shown in Table 4.3. CN is defined as nystagmus present at birth or noted in early infancy when fixation efforts develop. If CN coexists with a primary visual abnormality, it may be impossible to distinguish the nystagmus from the acquired type secondary to progressive visual loss. Only a reliable history that nystagmus was present in early infancy, or the opposite observation that the nys-

Table 4.3
Nystagmus of Infancy and Early Childhood

A. CN
B. Latent nystagmus
1. Vera
2. Manifest
3. Superimposed (two types)
C. Acquired nystagmus
1. Secondary to progressive visual loss
2. Spasmus nutans
3. Structural posterior fossa disease

tagmus initially developed after vision had deteriorated, can provide the necessary distinguishing information. Both appear to share the same variety of wave forms when examined with oculography. Most of these wave forms have never been reported as occurring in adults with acquired nystagmus.[16]

CN has certain clinically detectable characteristics, the presence or absence of which could establish or deny a congenital etiology. The nystagmus is binocular and associated (equal in the two eyes); it is never dissociated in direction or frequency and is rarely more than minimally dissociated in amplitude. CN is univectorial, that is, when horizontal (as it usually is), it does not change planes with gaze shifts as does acquired nystagmus, which is often horizontal in lateral gaze and vertical on upward gaze. Convergence damps several varieties of nystagmus but not as dramatically and consistently as with CN. The oft-quoted observation that CN stops behind closed eyelids and persists with eyes opened in the darkness is based upon the previously described role of fixation effort in the nystagmus generation; fixation attempt would be increased in darkness and eliminated with eye closure. Oscillopsia is rare, and head tremor is frequent, particularly as visual intent increases. Inversion of the optokinetic reflex[3] is diagnostic of CN, as is superimposition of a latent component (discussed below).

Latent nystagmus may be divided into three different types. *Latent vera* is a congenital form of nystagmus not present during binocular viewing but which appears in both eyes when one is covered. The fast phase always beats towards the viewing eye or, as demonstrated by van Vliet,[19] the eye which the subject believes is viewing. The shape of the slow phase, unlike CN but like the previously described gaze-evoked nystagmus secondary to defective tonic innervation, is a decreasing-velocity exponential (Figure 4.6). *Manifest latent nystagmus* is invariably diagnosed clinically as CN. Many patients referred to us with CN and strabismus are found, upon recording, to have the latent nystagmus wave form (Figure 4.6). The nystagmus is manifest because strabismus induces monocular viewing. *Superimposed latent nystagmus* is of two varieties. The first is a feature of classical congenital nystagmus wherein the neutral zone can be shifted by closing an eye (Figure 4.7). The nystagmus, before and after the monocular occlusion, has a "congenital" rather than a "latent" wave form; the latent superimposition is simply the shift. The second type, described by Anderson[20] and Kornhuber,[21] occurs in patients with rather nonspecific bilateral gaze-evoked horizontal-rotary nystagmus and often with primary position pendular rotary nystagmus. This nystagmus has no distinguishing features of CN except that monocular occlusion produces a typical latent nystagmus pattern. This "superimposition" establishes the ongoing nystagmus as congenital.

All forms of latent nystagmus have a high incidence of alternating hyperphoria.[20]

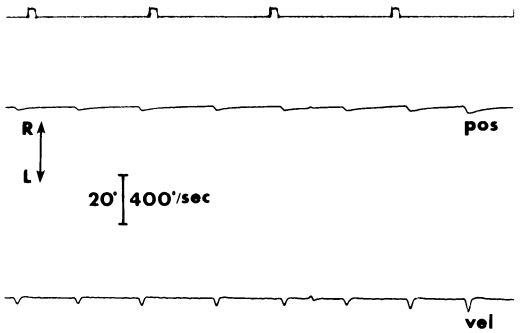

Figure 4.6. Oculographic tracing of latent nystagmus. The decreasing-velocity exponential slow phase is identical in latent vera and manifest latent nystagmus. *R*, right; *L*, left; *pos*, eye position; *vel*, eye velocity.

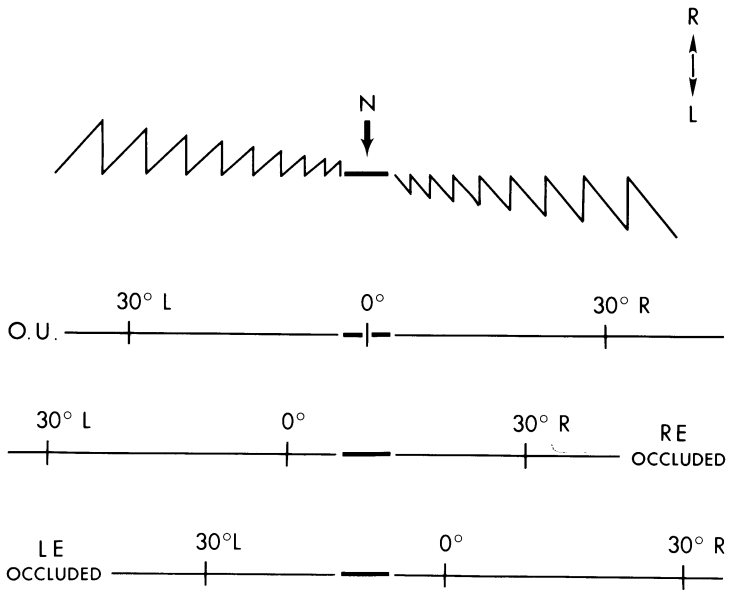

Figure 4.7. Depiction of the shifts of the neutral zone (*N*) in congenital nystagmus. An electronystagmographic tracing demonstrates an idealized nystagmus pattern with both eyes open (*O.U.*). The neutral zone extends over several degrees on either side of 0 degrees. When gaze is directed laterally, nystagmus of increasing amplitude develops with the fast phase in the direction of gaze. Occlusion of the right eye (*RE*) shifts the zone to the right; at 0 degrees there is left-beating nystagmus. Occlusion of the left eye (*LE*) shifts the zone to the left; at 0 degrees there is right-beating nystagmus. *R*, right; *L*, left. (Reprinted with permission from: R. B. Daroff and L. F. Dell'Osso: *Canadian Journal of Otolaryngology, 3*: 367, 1974).

Spasmus nutans is a rare constellation of head nodding, ocular oscillation, and torticollis. Our limited personal experience with this precludes insightful comment.

Posterior fossa disease in infants and children, as with adults, produces nystagmus. Unfortunately for the clinician, such nystagmus is quite variable and often lacks stereotyped features of the specific nystagmus patterns in adults. The developing nervous system reacts to dysfunction in an unpredictable manner.

Physiological End Point Nystagmus

Three basic types of nystagmus regarded as normal phenomena (physiological) can be described. These are: fatigue nystagmus, unsustained end point nystagmus, and sustained end point nystagmus.

1. Fatigue nystagmus, as the name implies, begins during extended maintenance of an extreme gaze deviation. Bárány[22] found it in 60% of normals with maximally deviated gaze exceeding 30 seconds. Nylen[23] described fatigue nystagmus as becoming increasingly rotary, with more prolonged and extreme deviation. Schmidt and Kommerell[24] studied six subjects and noted one with fatigue nystagmus; it began after a latency of 90 seconds. The nystagmus, bilaterally symmetrical and jerk in type, was of considerably greater amplitude in the adducting eye. The abducting eye manifested only irregular small amplitude and, often, pendular movement. Fatigue nystagmus is not a clinically important phenomenon since eccentric gaze is not maintained in routine examinations. Myasthenic nystagmus[25] is probably a pathological exaggeration of fatigue nystagmus.

2. Unsustained end point nystagmus is perhaps the most frequently encountered physiological nystagmus, but its incidence or characteristics have never been studied in a quantitative fashion. All experienced

clinicians recognize a few beats of nystagmus as being perfectly within normal limits at gaze deviations of 30 degrees or more.

3. Sustained end point nystagmus begins immediately or within several seconds after the eyes reach an eccentric lateral gaze position. Blomberg[26] studied 150 normal subjects at 40 degree horizontal deviation and claimed that none had nystagmus. Godde-Jolly et al.[27] found gaze-evoked nystagmus in 164 (63%) of 260 normal subjects, the vast majority of whom had an onset at greater than 40 degrees. Uemura et al.[28] stated with admirable honesty, "Distinct nystagmus which appears with a lateral gaze of 30 degrees or less from the midline is empirically believed to be pathological. The distinction between physiological and pathological gaze nystagmus is, however, arbitrary, sometimes difficult and may be impossible.... Only discrete nystagmus with a sufficiently large amplitude which is easily identified can be said to be unequivocally pathological."

Prior to our recent report on this subject,[29] the only previously published study was by Schmidt and Kommerell,[24] who analyzed five patients. The nystagmus, in illuminated conditions, varied from 1 to 3 Hz with an amplitude of 1–3 degrees and was the same with gaze to either horizontal direction. One subject had convergence jerk nystagmus, but the other four had conjugate jerk nystagmus with amplitudes varying between the two eyes. During maintained deviation in darkness, one subject manifested a high-amplitude pendular oscillation in the abducting eye associated with pupillary miosis and 4–5 diopters of lens accommodation. Another subject in darkness had equal nystagmus amplitude in the two eyes, whereas when fixating an illuminated target, the nystagmus was predominately in the abducting eye. A major finding by these authors was distinct variability within subjects recorded at different times.

We studied 11 normal subjects at 20, 30, 35, and 40 degrees of horizontal deviation.[29] Our sensitive (100 Hz) infrared recording technique did not permit measurement at more extreme gaze angles. Four subjects failed to develop nystagmus despite sustaining 40 degree deviations from 4 to 5 minutes. Six subjects developed sustained gaze-evoked nystagmus; in one, the nystagmus (0.5 to 1 degree and 1.0 Hz) began with only a 20 degree deviation. Only one subject developed "fatigue" nystagmus; it began after more than 4 minutes of deviation. The shape of the slow phase, in all instances, was linear.

Clinically Identifiable Nystagmus and Other Oscillations

Without the aid of eye movement recordings, most ocular oscillations can be recognized, and appropriate clinical inferences can be made. These have been described in more detail elsewhere,[3, 30] where specific references for each item are available. We have mentioned the major characteristics of congenital, latent, and bilateral horizontal gaze-evoked nystagmus.

Seesaw is a conjugate, pendular, torsional nystagmus with a superimposed dissociated vertical vector such that the intorting eye rises while the extorting eye falls. This nystagmus is usually associated with a bitemporal hemianopsia and a large parasella tumor.

Convergence-retraction nystagmus is associated with pareses on upward gaze and indicates a dorsal midbrain lesion.

Downbeat and periodic alternating have the same anatomical implications. They reflect intrinsic brainstem disease but should particularly alert the clinician to extra-axial compression of the craniocervical junction, such as an Arnold-Chiari malformation.

Upbeat nystagmus is secondary to an extensive lesion of either the cerebellar vermis or the medulla.[31]

Ocular myoclonus, a pendular vertical nystagmus synchronous with a palatal oscillation, results from pseudohypertrophy of the inferior olivary nucleus. The later occurs following lesions in the contralateral dentate nucleus of the cerebellum or ipsilateral central tegmental tract.

Ocular bobbing describes rapid, conjugate, fast downward jerks of the eyes fol-

lowed by a slow return to mid-position. The clinical setting is usually a comatose patient with total paralysis of spontaneous and reflex horizontal eye movements. The usual cause is a massive pontine lesion but extrapontine compression, obstructive hydrocephalus, or metabolic encephalopathy are occasionally responsible.

Superior oblique myokymia denotes extremely rapid, small uniocular torsional movements which are usually intermittent. The patient experiences oscillopsia but usually has no underlying discernible neurological disease.

Positional nystagmus (synonymous with "positional vertigo," the invariably associated symptom) of the peripheral variety[8, 9] occurs while the patient is in the supine position with the head straight or turned to either side. The onset is after a brief latency and not immediately upon attaining the posture. If the position is maintained, the nystagmus will gradually fatigue and disappear. The patient can then be seated and challenged again by the critical position where the nystagmus will be less intense and prolonged than previously. With repetitive challenges, the nystagmus will habituate entirely. Another important feature of this type of positional nystagmus is its lack of reproducibility, despite a history of vertigo typical of the condition.

Nystagmus secondary to *extra-axial brainstem compression*, such as an acoustic neuroma or cerebellar mass, is often bilateral gaze-evoked horizontal with large amplitude and lower frequency toward the side of the compression and high-frequency, small amplitude jerks to the opposite side.

A number of so-called "cerebellar eye signs" follow:

Rebound nystagmus is of two types. The clinically recognizable form is a gaze-evoked lateral nystagmus which, upon return to primary position, beats transiently in the opposite direction.

Ocular dysmetria occurs following a saccadic refixation in which the eyes oscillate transiently before coming to rest. Eye movement recordings demonstrate that the interval between the initial saccade and each of the following saccades is approximately 200 milliseconds. The number of saccades required for final target foveation is dependent upon the gain and varies from one to many.

Ocular flutter is a momentary rapid binocular conjugate horizontal oscillation occurring during straight-ahead gaze. Eye movement recordings demonstrate that the saccades have no interval between them. Flutter may appear following a saccade. This we call flutter dysmetria. It is clinically indistinguishable from classical ocular dysmetria, but oculography discloses the absence of latency between the oppositely directed saccades which constitute flutter. A single such return saccade following a saccadic eye refixation is called "dynamic overshoot" and occurs in normals. Bursts of flutter are indistinguishable from voluntary nystagmus, which also consists of oppositely directly no-latency saccades.

Opsoclonus is the most dramatic of the oscillations. These are conjugate multivectorial (except rotary), unpredictable, chaotic saccadic eye movements. In infants with cerebellar ataxia and extremity myoclonus, a nonmetastatic complication of an otherwise occult neuroblastoma must be ruled out. In the absence of a neuroblastoma, ACTH is often an effective therapy. Opsoclonus may also be associated with a benign self-limited postinfectious encephalopathy and, in older patients, a remote nonmetastatic complication of carcinoma.

Macro-square wave jerks are large amplitude, intermittent, to-and-fro horizontal saccades with the intended fixation point at one end of the oscillation. This is usually associated with cerebellar outflow (dentato-rubral) disease.

Macrosaccadic oscillations resemble the above but straddle the fixation point.

ADDENDUM

After submission of this manuscript, we added Table 4.4, which categorizes the major types of jerk nystagmus on the basis of the slow-phase shape. As mentioned ear-

Table 4.4
Classification of Jerk Nystagmus by Slow-Phase Shape

Linear
Optokinetic
Vestibular
End point (<40°)
Fatigue
Myasthenic
Drug
"Pursuit"
Gaze-evoked "linear"
Decreasing-velocity exponential
Gaze-paretic
Latent
Manifest latent
Rebound
INO Abduction
End point (>40°)
Increasing-velocity exponential
Congenital

lier in the text, the shapes are of three types: linear, decreasing-velocity exponential (Figure 4.3), and increasing-velocity exponential (Figure 4.2).

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