Paparella: Volume II: Otology and Neuro-Otology

Section 1: Diagnosis of Disorders of the Ear

Chapter 3: Vestibular Physiology:

Its Clinical Application In Understanding the Dizzy Patient

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This chapter is designed and written for the clinical practitioner who is at times bewildered by the dizzy patient and the multitude of causes that this problem might represent. It does not purport to inform the reader in depth concerning what is known of vestibular neurophysiology today, but it hopes to give him a qualitative conceptualization of vestibular mechanisms in health and disease and, to the degree it does so, make the patient with such a problem an interesting challenge. The dizzy patient may be a perplexing diagnostic problem because he presents himself very often for treatment without any objective sign or test result of diagnostic value. Even audiometric data may be absent or non-revealing. A thorough and practical knowledge of how the vestibular system works as it relates to the symptoms of vertigo may be then the physician's best diagnostic tool.

Some initial points need to be made for this particular appreciation of vestibular function in understanding the dizzy patient.

The first is that all that is dizzy is not vestibular. Many body systems can produce dizziness - the cardiovascular system, the extravestibular tracts, the cardiopulmonary system, the metabolic system, the oculomotor system, the hormonal system, etc. Herein lies the first big fork in the flowsheet to diagnosis - the distinction between nonvestibular dizziness (an altered sensation of awareness of environment or self) and vestibular dizziness (vertigo or a specific alteration of orientation which involves motion of either the subject or his environment).

The second point is that a knowledge of the minute structure of the end-organ is not clinically essential. It can be simplified in a conceptual manner for the purpose of understanding the dizzy patient. The endolymphatic system consists of the coiling cochlear duct at one end, the three semicircular ducts on the other, and three specializations of this continuous tube in between - the utricle, the saccule, and the endolymphatic duct and sac. Any movement of the head in which there is some angular acceleration causes a piling up of endolymph on one side of the cupulae of two or more of the six semicircular canals (which are orthogonally paired structures), and the brain is signaled. The density of the cupula and endolymph is probably the same, since their refractive indices are the same. Hence, gravity cannot affect the cupulae. The utricle and the saccule contain flat sensory areas, the maculae, overlain by a gelatinous coat studded with calcified bodies, the otoliths. These organs seem best equipped to sense linear acceleration, of which gravity is one variety.

The third important point is that the vestibular end-organs are dynamic structures. They are dynamic in three ways:

1. They respond to linear and radial accelerations.

2. They are not silent until stimulated, but constantly discharge a resting pattern of signals to the brain. Acceleration or a change in acceleration deviates the cupula and produces a change in this pattern of signals, and it is this change that is distributed to the brain and interpreted.

3. There are two sets of vestibular systems, each constantly signalling. A difference in the signal pattern between right and left is produced by an acceleration, and it is this difference that is the relevant quantity to the brain.

The Balance Theory

The vestibular system, being a very old system phylogenetically, has diffuse connections with the central nervous system. Some of the important areas upon which the vestibular system discharges are illustrated here, but the illustration is on a conceptual rather than on an anatomic basis. On normal stimulation there is brought about a change in impulse patterns of a precise and specific nature on the two sides. On one side the resting potential is modulated downward, and on the other side the resting discharge is modulated upward. As far as we known, the difference between the downward modulation on one side is precisely the same. Thus, the two sides of the brain are informed in an equal but opposite manner. This polarity is important in understanding the system; the equality is even more important. The numerous areas to which the vestibular system discharges respond in recognition. The cerebral cortex interprets the change as a movement of specific direction and speed. The eye muscle nuclei move the eyes compensatorily to retain the field of last gaze, opposite to the motion of the head, as a protective mechanism in retaining orientation to environment. The anterior horn cells in the spinal cord adjust trunk and limb muscles, and the cerebellum adjusts muscle tonus to meet the new situation. These processes are probably partly instinctual and partly learned. The cortex, for example, likely underwent a prolonged period of training while the organism was learning to stand and walk. It learned to match a certain degree of cupular displacement in response to sudden head motion to a specific degree of alteration of the surrounding environment by matching a number of modalities against it - for example, the eyes informed the brain that the body has moved so far, and tactile modalities informed the brain when the organism had hit the floor. Over the years, then, the brain had learned exactly what to expect from its vestibular end-organs in combination with other modalities. Being a superb computer, by childhood the brain has integrated these sensations to a high degree.

Again it is important to conceive of the vestibular apparatus as two systems, right and left, in constant dynamic balance, one checking against the other, working as a team to inform the organism of movements and head positions, and adjusting the body to meet these new conditions.

Last, it is important to conceive of each vestibular system, right and left, as a complex system from end-organ to cortex, and not as an isolated end-organ on one side.

Disease Strikes

When there occurs a sudden pathologic diminution of function of one vestibular system (and vestibular crises are usually of the diminution or destructive variety rather than the irritative variety), as for example, a Ménière's speel of one end-organ, there exists a major imbalance. The involved side is no longer able to deliver its equal and opposite fund of information to the brain. The two systems are discharging at rest at an unequal intensity, and unequal intensity of discharges has a specific meaning to the brain. The sequelae of this imbalance are manifestations of a relative hyperfunction of the intact side; thus uncontrolled and prolonged vestibular reflexes result.

The disparate message arrives at the cerebral cortex, and the cortex interprets this unbalanced information from two sides in the only way it can: in the light of past experience. The cortex interprets it as a condition of constant motion - and this is our definition of vertigo. This misinterpretation of the actual state of affairs is a rotary sensation when the whole end-organ is involved because the six semicircular canals predominate in their overall effects over misinformation from the four otolith organs alone, simply by the law of mass action. Thus the sensation is of a rotary nature. It may be of a pitching, yawing, or rolling character, but always of a rotational nature because of this predominance of innervation.

The same massive imbalance in discharges arrives at the eye muscle nuclei and the reticular formation. The imbalance, interpreted as before in the light of past information and training, directs the eye muscle nuclei to deviate the eyes in the direction of last gaze to retain orientation; the slow component of nystagmus is born. The eyes, however, cannot continue to tract indefinitely in any single direction because of their anatomic limitations, so after a deviation specific to the number of motoneurons has transpired, inhibitor neurons in the reticular formation cut off the incoming flow from the vestibular nuclei and, at the same time, reticular activating neurons direct the ocular muscle nuclei to return the eyeballs to the point of gaze at which the slow component began the deviation (across the midline). This second phase of eye deviation is a much faster one because it is a compensatory or recovery phase. The quick component of nystagmus is thus generated. The reticular activating neuron, having fired, enters into its refractory period, and the end-organ inflow from the vestibular nuclei resumes its effect upon the eye muscle tracts - the eyeballs are directed again to retain the field of last gaze. This repetitive attempt to retain the last field of gaze by a conjugate movement of the eyes and a rapid reflex return of the eyeballs across the midline in compensation is our definition of vestibular nystagmus.

The same imbalance of information is transmitted from the vestibular nuclei down the spinal cord to anterior horn cells, instructing the postural and locomotor muscles to meet a new situation that never comes; staggering and ataxia results.

The imbalance in impulses also plays upon the dorsal efferent nucleus of X. At first this nucleus effects only a cessation of peristalsis. Gut activity is not needed in an emergent situation. If the imbalance is massive and continuous, however, this nucleus is heavily stimulated, and reverse peristalsis occurs, with resultant nausea and vomiting.

The effects mediated by the cerebellum in response to a massive imbalance on the two sides are only beginning to be understood. In a matter of minutes the cerebellum imposes a virtual shutdown of electrical activity of the vestibular nuclei (at least the medial nucleus, the major way station for incoming canal impulses) by virtue of its profound inhibitory influence on vestibular activity. The cerebellum does not eliminate the great imbalance by this shutdown because not all information from the end-organs distributes to the brain through these nuclei - some fibers from the end-organ distribute straight to numerous other parts of the brain stem and cerebellum without vestibular nuclear synapse. The nuclear shutdown does not then eliminate the problem, but it does serve to render the imbalance at a lower level of magnitude so that the full effects of the imbalance do not distribute through all available vestibular pathways.

The Physiology of Repair and Compensation

The organism then sets about trying to restore the situation. It cannot long endure it; the organism will eventually die through dehydration and fluid and electrolyte imbalance. Restoration of equilibrium between the two centers brings about resolution of the uncontrolled reflexes. This can be done in three ways: (1) restoration to health of the diseases system, which may take hours to days; (2) central suppression of the intact side by evocation of inhibitory tracts in the central nervous system; and (3) generation of a new electrical activity in the underdischarging system to balance the normal but now relatively hyperactive side. These include probably all theoretical mechanisms involved. In practice it is very likely that all three mechanisms go on at once in varying degrees. For example, in the crisis of Ménière's disease the end-organ heals itself in a few hours, and a normal or near-normal discharge pattern from the end-organ resumes. The cerebellar "clamp" is not needed, or at least only temporarily. Reflexes then revert to normal as equal and opposite reactions are signaled from the two end-organs.

Another example would be acute suppurative labyrinthitis. In this disease the end-organ is destroyed and, since it cannot rebuilt itself, restoration must be a central process. Very quickly the cerebellum imposes vestibular and nuclear shutdown, and this provides a barely tolerable situation for the organism as long as there is no or at least minimal stimulation of the opposite end-organ to accentuate the great imbalance. For this reason patients in vestibular crisis remain perfectly still with as little head motion as possible. Motion of the head results in accentuation of the imbalance, with waves of vertigo and vegetative symptoms. Then, over a matter of days and possibly weeks, a new resting electrical activity is generated in the denervated vestibular nuclei. As this new activity builds, symptoms begin to abate and the cerebellar shutdown is slowly released. When the activity is full and matches the other side, symptoms disappear except for varying degrees of motion intolerance. Motion interpretation involves integration, and this must gradually be built up following regeneration of resting activity in the nuclei.

We do not know what stimulates the generation of the new resting activity in the denervated nuclei, but we know of certain essentials for it. Certainly there must be a chronic vestibular input imbalance or, more simply, a vacuum stimulating its need. The speed at which it is brought about is dependent on the severity of the imbalance stimulating it, and the ability of the central nervous system (CNS) to respond. This ability is a function of the vigor of the whole organism - age of the patient, availability of neuron arcs, efficiency of the central nervous system vascular supply, and so forth.

Clinical Applications of the Balance Theory

From such a consideration of the balance theory of vestibular function, we arrive at two axioms:

1. In vestibular crises of any severity, there will always be labyrinthine nystagmus.

2. If the symptoms last continuously for more than two or three weeks, the cause is not vestibular.

These axioms can be applied clinically. The first can be helpful if the patient, while dizzy, can be observed by the physician, or, indeed, any instructed person. *If the patient in a significant speel does not have spontaneous labyrinthine nystagmus, the disease is not vestibular.* The physician may not often have opportunity to observe a spell because the patient presents usually between spells. However, the patient's spouse can often-times be a surprisingly good observer once instructed. The physician can instruct the spouse in the office at the initial visit by pointing out carefully the features of the nystagmus produced by the simple caloric test that he performs in the course of his workup. The author has come across some people who have become surprisingly astute observers after little instruction. This may seem at first self-evident, but it is an important starting point because it may clearly establish the disease as vestibular or extravestibular early in the diagnostic workup. If the otolaryngologist is able to establish its nature as non-vestibular, this alone is helpful to the referring physician. If he can establish the disease as vestibular, he can proceed to the next important fork in the flowsheet, that of central or peripheral causation.

The second axiom is also helpful in this regard. If on close questioning the patient states that his dizziness has been nonepisodic and continuous for, say, two or three months, then his disease cannot be vestibular. As we have previously pointed out, the vestibular system does not work in this way. There are virtually no clinical exceptions to this axiom.

Vestibular Function Tests - What We Can Learn From Them

The goal of vestibular function tests in the present state of the art should be primarily to distinguish a vestibular disease as either end-organ or central. If this can be determined with satisfaction, this alone will be a laudable achievement. It is frequently an immense relief to a patient to be told that his disease is end-organ and that, whatever follows in the way of

symptomatology, his condition will not shorten his life by one day. Even if his disease is not directly treatable, he can at least be assured of eventual relief. If, on the other hand, the disease can be recognized as central, the patient can be put in the hands of the proper specialist until it is diagnosed or time and the emergence of new symptoms make it diagnosable.

Rotation Tests

Rotation tests are the oldest of vestibular tests and have advantages and disadvantages. The major advantage is that cupula deviation can be produced to a precise degree, but this takes sophisticated and expensive equipment. The major disadvantage is that it stimulates both ears at once and gives no laterality information. It should not be considered purely a research test, however, because some clear and useful information can at times be adduced. If, for example, a gross difference (more than 30 per cent) in nystagmus duration is produced by equal spins in the two directions, there either *is* or *has been* a significant vestibular incident. This is not specific, but it helps one along the flowsheet to diagnosis. This rotation test can be done rather simply by putting the patient in any chair that will rotate, turning the patient up by hand 10 turns in 10 seconds, and then stopping him suddenly. After a few minutes, the turns can be repeated in the opposite direction.

Another example of the utility of this test is in the very young child brought in by his parents because of apparent severe deafness. Very young children require special audiometric equipment and skills for hearing quantification, such as electroencephalographic audiometry, available for the most part only in large medical centers. The child can be positioned on the lap of a parent, who holds the head of the child under his own chin so that the heads are on nearly the same axis. The rotation test is done, and if the child's nystagmus is grossly less than the parent's, particularly if these is little or no nystagmus, it can be accepted that there is a severe derangement of the inner ears; immediate and extensive referral is warranted. Although there may seem to be little urgency, experts in education of the deaf are placing hearing aids on children at an increasingly early age.

Electrical Tests

In this category we have only the faradic and galvanic variety, with the electrode placed over the mastoid cortex. This produces a depolarization of the vestibular nerve and precipitates gross movement of the body. The electrical charge necessary to penetrate the skin and temporal bone to the nerve is so large that it is a painful and, thus, impractical test. Some form of electrical stimulus will be the test of the future, however, for it is precisely quantifiable in terms of strength. Some future investigator will give us an electrical stimulus that can be driven without significant pain, much like square wave testing of the facial nerve.

Postural Tests

Postural tests are performed for the detection of positional nystagmus. They can be performed with and without visual fixation. With visual fixation the patient fixes his eyes at a

point in each test position with the eyes in cardinal position of gaze. Testing without visual fixation is done with Frenzel glasses, or preferably by electronystagmography. The usual test positions are (1) upright with head erect, (2) recumbent with left ear down, (3) recumbent with right ear down, and (4) head hanging, with vertex pointed at the floor. Whether the head is moved with the body or the head moved on the body with neck torsion is not important unless one is seeking to distinguish between gravity-actuated nystagmus and nystagmus produced by vascular embarrassment or from torsion of neck vessels.

The important features of the provoked nystagmus to note are latency, fatigability, direction-changing or direction-fixed, and perversion. No latency, or a very short one, is indicative of a central lesion; a long latency is indicative of a peripheral lesion. In the latter case, the latency may be as long as 15 to 20 seconds; hence, the position must be held for that period in search of nystagmus. A nonfatigable nystagmus (that lasting as long as the position is held) is indicative of a central lesion; fatigable nystagmus is indicative of a peripheral lesion.

Direction-changing nystagmus is nystagmus which changes in direction from one head position to another, or one examination to another (Aschan type II). Direction-fixed nystagmus is in the same direction regardless of position or time (Aschan type I). Direction-fixed positional nystagmus is virtually always end-organ in origin. The lesions in the majority of patients with direction-changing nystagmus will also be located in an end-organ, but a significantly high percentage will be central in origin. For that reason, a direction-changing positional nystagmus should be a red flag of warning.

Perverted nystagmus is defined as that which is unexpected in terms of the stimulus. For example, a rotatory nystagmus upon stimulation of the horizontal semicircular canal would be a perverted one. Positional nystagmus of the end-organ variety produces a horizontal-rotatory nystagmus and, less often, a pure rotatory or pure horizontal nystagmus. Thus, a vertical nystagmus on positional testing would be a perverted one. Perverted nystagmus almost always means a central lesion.

Postural vertigo is a disease characterized by positional nystagmus. Repeated examinations may be necessary to elicit the nystagmus, but they are essential to a definitive diagnosis. There is a specific and relatively common form of this disorder termed "benign paroxysmal postural vertigo" (Aschan type III). With the involved ear undermost, after a definite latent period there appears a rapid labyrinthine nystagmus of brief duration (fatigable) accompanied by intense vertigo. It is direction-fixed. It is thought to be of peripheral origin, but this is not known with certainty. Physiologically it appears to be an otolithic defect, with consequent loss of otolith-organ governing or modulating influence over the semicircular canals. It is a self-limited disease.

Caloric Testing

There are a great variety of caloric tests used clinically; they vary essentially only in volume and temperature of water used. The important features of caloric testing of the labyrinth are twofold: (1) enough stimulus should be provided to compare the two sides, and (2) enough

stimulus should be provided to evoke all the sequelae of cupula deflection. Unless these are done, much valuable information may be missed. Threshold tests are of limited value, as they provide only part of the picture. A suitable screening caloric examination is the injection of 5 mL of ice water over a 5-second period directed at the posterosuperior quadrant of the tympanic membrane under direct vision. If this does not drive each labyrinth to the desired degree, as indicated previously, 10 mL of ice water may be used, or, if this is not adequate, 20 mL of ice water. If even this does not produce nystagmus or vertigo, that vestibular apparatus is said to be inactive.

The important initial questions to be answered by the caloric tests are (1) Does the labyrinth work or not? and (2) Does the caloric qualitatively (not quantitatively) reproduce the patient's spell?

At a minimum the above information should be provided by the caloric test. By following these principles, the discriminating observer can, at times, adduce significant information that may pinpoint the lesion. If enough stimulation is given to evoke all the sequelae of cupula deflection routinely, a missing component may be highly significant. For example, if there is nystagmus and vertigo but no nausea upon adequate stimulation, this may be indicative of a low pontine or medullary lesion with a cutoff of impulses to the dorsal efferent nucleus of X. If there is nystagmus and nausea but no vertigo, this may indicate a midbrain lesion with a cutoff of impulses to higher centers, where vertigo is interpreted. If there is vertigo and nausea but no nystagmus, this may indicate a median brain stem lesion with cutoff of impulses to eye muscle nuclei, such as in syringobulbia or multiple sclerosis.

Electronystagmography

The technique of electronystagmography (ENG) and interpretation of electronystagmogram is not within the purview of this chapter (see Chap. 4 for detailed discussion of this technique), but some comments must be made upon this relatively new study for the sake of completeness.

There is no question that ENG has moved out of the research laboratory and is now properly a clinical neurotological test. It allows us to determine the intensity of vestibular response (eyeball speed in the slow component), to detect latent nystagmus (that present with the eyes closed, or the eyes open in the dark), and to detect the gaze nystagmus characteristic of some central lesions and late peripheral lesions. It allows us to perform positional tests with a higher degree of yield, since optic fixation can be eliminated, and it provides us with a permanent record for analysis and comparison. Finally, it allows us to observe some of the provoked features of nystagmus, such as secondary phase nystagmus (or provocation nystagmus), which might otherwise be missed.

The neuro-otologic diagnostician can today in the vast majority of patients determine whether the lesion is vestibular or extravestibular. This can be accomplished by a careful consideration of the history and application of the tests described previously, to which is added the complete audiologic battery and appropriate radiographic studies. The dilemma of the diagnostician at this point is determination of a central versus a peripheral lesion. It is well

known that the majority of vestibular lesions are peripheral, but this knowledge does not help us in the individual patient. ENG may be helpful in this regard, and indeed may give us the sole clue as to the central nature of the lesion. The following four steps should be features of every routine ENG in order to increase its diagnostic yield.

Calibration Overshoot. Calibration overshoot is the appearance of spikes on the leading corners of the square waves produced on the strip chart during the calibration exercise. These must be on the leading corners of either the left or the right eye track in order to be significant. Spikes appearing in the center of the top or bottom of the square wave or near the trailing edge are not significant. Calibration overshoot appearing in 50 per cent or more of the square waves is indicative of cerebellar disease. Simmons has shown an 83 per cent correlation between this sign and cerebellar disease, an extremely high correlation for ENG. It is a manifestation of ocular dysmetria. It is a very attractive feature of ENG not only because of its significance but in that it adds no time to the study.

Abnormal Pendulum Tracking. A pendulum can be as simple an arrangement as an orange-painted golf ball suspended from the ceiling by a string. The ball is set in motion and the patient instructed to follow the pendulum with both eyes open but one eye covered. It is extremely important that the tracking be recorded monocularly. If both eyes are allowed to tract together, the normal eye will harness the abnormal eye, and these important signs may be missed. Corvera has the most experience in this regard and describes three types of tracking response that can be very useful. Normally and in some patients with peripheral lesions a very clean sine wave of slowly decreasing amplitude is demonstrated. A second type is a wiggly wave superimposed on the sine wave, but wherein the sine wave is still clearly discernible. This is an indication of peripheral disease. The third type is that in which the sine wave is indistinguishable, indicating a central lesion. It is another measure of ocular dysmetria.

Failure of Eyes-Open Damping. This is frequently referred to in the literature as a positive *ocular fixation index.* At the height of the caloric nystagmus run or just past it, the patient is instructed to open the eyes and fix on an object in the cardinal position of gaze. Normally and in patients with peripheral lesions the speed of the slow component will be significantly diminished by fixation in the light, and on eye closure the slow component speed will increase. When the eye speed does not decrease, and particularly when it increases over that with the eyes closed, a central lesion is indicated. The ocular fixation index is derived by dividing the eyes-open slow component speed by the eyes-closed slow component speed. When this figure is 0.8 or 0.9, it is highly suspicious of a central lesion, and when it is 1.0 or greater it is virtually diagnostic.

Perstimulatory Fatigue. This phenomenon has been termed *decruitment* by Torok. We elicit this phenomenon by stimulating the ear first with 5.0 mL of ice water and 5 minutes later by 10 mL of ice water. The second stimulus normally produces a greater slow component speed than the first because the vestibular system, like all sensory systems, produces a straight-line response in terms of intensity function. If the second response is significantly less than the first, fatigue or decruitment is said to be present. This is a manifestation of a depleted number of

neurons in the reflex arc and indicates a central lesion. It is very important that adequate alerting be performed, especially during the second stimulation. As a double check, if the second stimulation response is less than the first, the alerting mechanism should be switched from a mathematical to the conversational type.