Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 4: Inner Ear

Chapter 48: Differential Diagnosis of Dizziness and Vertigo

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When dizziness is a complaint presented by a patient to the physician, it indicates that the patient is attempting to describe his or her disturbed relationship to space. Subjective orientation of the body in space is controlled by a complicated system that includes the following:

1. The eyes and eye muscles.

2. The proprioceptive system.

3. The labyrinth (semicircular canals and utricle).

4. Cranial nerve VIII.

5. The vestibular nuclei in the brain stem.

6. The neural pathways from the vestibular nuclei to the midbrain and to the temporal lobe (medial longitudinal fasciculus).

7. The central coordinating mechanism in the cerebellum.

8. The vestibulospinal tract (posterior columns).

9. The efferent fibers coming from muscles and joints to the posterior columns.

The end-organ of equilibrium (semicircular canals and utricle), vestibular division of cranial nerve VIII, and vestibular nuclei are commonly referred to as the statokinetic system.

Disturbed function of any of the parts mentioned may produce dizziness. *However, only a disturbance of the statokinetic system gives rise to true, whirling vertigo;* this somewhat dogmatic statement implies that dizziness may mean several sensations to the patient. The patient may experience true whirling, often in a given direction, or the subjective sensation may be one of uncertainty, insecurity, giddiness, weakness, confusion, blankness, or unsteadiness. It may also include light headedness or near syncope. The complaint may be "tipping" or "propulsion". Whirling or spinning sensations, or a subjective feeling of propulsion or tilting, direct attention to the statokinetic system, whereas the other, less specific complaints may be caused by disturbance in any part of the body.

When the three primary systems that control equilibrium (eyes and eye muscles, statokinetic system, and proprioceptive system) are intact, our relationship to space is properly oriented (by experience, learning, and reflex activity) and we are stable. If one of these systems is reacting abnormally as a result of disease or trauma, or if one of them is not functioning, the equilibrium is disturbed. If two of the systems are affected, a patient is usually incapacitated and unable to maintain stability at all. Serious end-organ disease alone is incapacitating.

Of all symptoms a patient presents to a physician, dizziness is one of the most common as well as one of the most confusing. Within certain limits, it is possible to classify dizziness according to recognizable entities. By so doing, the physician not only clarifies part of the confusion, but also establishes some basis for rational therapy.

The statements "within certain limits" and "clarifies part of the confusion" are used advisedly, since, regardless of the classification, *there will always remain certain patients whose dizziness is never exactly understood* and whose treatment must be a regimen of trial and error. This does not imply that classification, so far as it is possible, is of no value. Just the opposite is true. The physician must have certain established symptom complexes in mind, which, when all the criteria of each are met, point to a specific diagnosis and lead to a specific treatment.

History

The first step in evaluating a complaint of dizziness is the recording of a detailed history. *More insight into the patient's complaint can be gained from a carefully taken history than from any other source.*

What does the physician with to learn from the history? *First, he or she hopes to determine whether the patient has experienced true whirling, or propulsion, or falling.* The direction and character of motion are of little value as compared with the fact that there is a sensation of motion. A question frequently asked of the patient is, "Does the room move around you, or do you seem to be whirling?" An affirmative answer to either part of the question is sought. Too often the examining physician assumes that there must be a sense of motion if the patient is dizzy. A quick statement that "the room seems to move around" must not be accepted immediately but should be carefully probed until the examiner is convinced there is a sense of movement. There is a true sense of movement if, when the patient's eyes are open, objects seem to move around and if, when his eyes are closed, he feels that he is moving. Both conditions indicate vertigo and narrow the field of investigation to the end-organ, cranial nerve VIII, or vestibular nuclei and their central nervous system connections, if the premise that vertigo arises only from the statokinetic system is accepted. If a sense of motion cannot be proved by questioning, the entire body becomes the field for investigation.

Second, the physician is interested in any associated disturbance of hearing. If hearing loss or tinnitus, or both, accompany the dizziness, localization of the cause may be easier. The physician should expect to find disturbances of hearing associated more commonly with true whirling than with other, less well-defined sensations. However, he or she should be aware that true vertigo can occur without hearing loss.

Third, the physician wants to discover the pattern of the dizziness. The primary object of the questioning here is to determine whether the sensation is continuous or paroxysmal, followed by periods of complete relief. It is of considerable value to know whether the dizziness is brought on or made worse by changes of position and whether it is present only in certain positions. If a definite pattern can be worked out, the diagnosis is frequently established without further investigation of the entire body.

Fourth, the physician is interested in learning the degree of dizziness. Severe vertigo causing nausea and vomiting is, in the absence of acute disease of the central nervous system, most frequently due to an end-organ lesion. Less severe dizziness may arise from disease in any part of the body.

Examination

How much can the physical examination help the physician in evaluating the dizzy patient? Frequently the examination shows normal conditions. In a discussion of this length it is impossible to detail all the physical signs of the diseases in which dizziness may be one symptom. It is possible, however, to point out certain signs that can be easily checked and that are important in determining the probable cause of the dizziness.

The most important physical sign is spontaneous nystagmus, and the spontaneous nystagmus of greatest significance is in the primary position with the eyes straight forward. Nystagmus on lateral gaze is often physiologic and not of importance unless excessive or unusual in type. *Spontaneous vertical nystagmus or diagonal nystagmus* is believed to be diagnostic of central nervous system disease. *Nystagmus in a different direction in the two eyes* is always indicative of central nervous system disease. It is well to remember that the barbiturate drugs, used in excess or over long periods, may produce nystagmus. Fine nystagmus may be missed completely, since the normal fixation power of the eye muscles may be strong enough to overcome it. It is important, therefore, to cover the eyes with plus-20 lenses or to examine the fundi with an ophthalmoscope to determine the presence of fine nystagmus. The physician may not always see this nystagmus. But, *when he does, he may be sure there is disease* and reasonably sure the disease lies within the statokinetic system.

Gross muscle imbalance or simple glaucoma may constitute the entire cause of the dizziness. Therefore, a complete eye examination may be necessary. Tests of the corneal reflex should not be neglected, since a diminution in or loss of normal corneal reflex is often the first additional sign in tumors of the cerebellopontine angle, regardless of type. The possibility of acoustic neuroma should always be borne in mind. However, this is an *uncommon occurrence* when compared with other disturbances causing dizziness.

Examination of the ear is of particular value in attempting to discover the cause of dizziness. Impacted wax, foreign bodies in the external canal, retracted eardrums, and trauma of the middle ear may all be causes of mild vertigo. The presence of chronic suppurative otitis media should always be suspected of being the probable cause of the dizziness. A positive reaction to a fistula test when there is a perforated eardrum, even though the ear is dry, is evidence enough to indict the ear as the underlying cause of vertigo. A positive fistula reaction with an *intact membrane* may be seen in patients after a fenestration operation or

after stapedectomy or with central nervous system lues. These conditions are, fortunately, obvious and rarely misinterpreted.

The more important examination of the ear in evaluating the dizzy patient is the functional examination. The attempt here is to determine, as far as is possible with present equipment and present knowledge, the function of the cochlea, labyrinth, and cranial nerve VIII. An audiogram should always be done even though there is no complaint of hearing loss. Not infrequently high-tone losses will be discovered, which, if unilateral, may be of aid in localizing abnormal changes. Masking of the opposite ear when testing for bone conduction is a practice *that must be followed* if the physician is to interpret the type of hearing loss found. With a severely damaged nerve on one side, the patient may still hear loud bone-conducted stimulation in the good ear.

Tests with the tuning fork should be employed to support or refute any questionable audiometric observations. Here, again, masking with the Bárány noise box or other suitable sources of sound should be carried out when the physician is testing for bone conduction loss and also for air-conduction loss, if it is severe. *There are few characteristic audiograms of specific disease.*

In general, disturbances of equilibrium and diminution of cochlear function parallel each other when there are lesions of the end-organ and the nerve. In the brain stem, however, the vestibular and cochlear nuclei are separated widely enough so that *a minute lesion* may affect either without also involving the nuclei of other cranial nerves. Brain stem lesions may, therefore, give rise to vertigo without hearing loss.

The functional examination of the semicircular canal system is most easily done with the caloric test. Commonly employed is the Kobrak technique, which utilizes ice water or water at a given number of degrees below body temperature. Some investigators prefer to test with both warm and cold water. The determination of directional preponderance by the use of warm and cold water testing at fixed degrees above and below body temperature may elicit interesting information but does not often assist in arriving at a clinical diagnosis.

It is important that the physician remember to *use the same technique on all patients* under conditions that have as few variables as possible. Evaluation of patients and of opposite ears in the same patient is then possible. Ice water directed toward the posterior portion of the eardrum with the patient erect and the head tilted 30 degrees forward (to bring the lateral semicircular canal to the horizontal position) will primarily stimulate the anterior vertical canal. Similarly, if the patient is supine with the head tipped 60 degrees backward, the horizontal canal is maximally stimulated. However, all canals probably respond. Just above threshold response for the average person is the desired stimulus. This can be obtained best by a duration of stimulus of 8 to 12 seconds.

It is convenient to use a 22-gauge needle on a Luer-Lok syringe and to observe the stream of water through an ear speculum. Since the latent period of onset of nystagmus is determined by the strength of the stimulus, *the temperature of the water used should be constant.* The intensity of the reaction varies with the duration of the stimulus. Therefore, *the duration should be constant* from patient and from ear to ear in the same patient.

Since the reaction to caloric stimulation varies in different persons and also in the same person at different times, there is a wide range of reactions that must be considered normal. However, the average limits must constantly be remembered. The physician cannot expect detailed information from the usual office caloric test. He or she can be sure of *hypoactivity, hyperirritability, or complete loss of function.* Knowledge of these conditions is of value in classifying the probable underlying disease.

There has been controversy regarding whether any labyrinth is "hyperirritable". It has been stated that only normal or decreased responses are to be expected from caloric stimulation. However, almost 100 per cent of patients who have a proved history of *motion sickness* will react to caloric stimulation with definite overresponse. This response is not the subjective functional fear of the sensation of vertigo produced by caloric stimulation, which is often seen in patients with known anxiety tension states. The response in patients who have motion sickness is an actual and visible increase over the known limits of normal response. In other words the nystagmus, past-pointing, and falling are all severe, nausea and/or vomiting may be induced, and the subjective vertigo is violent. Whether the physician wishes to label this "hyperirritability" or a "low-threshold" labyrinth is unimportant. The important information gained from such a response is that this individual has (perhaps from birth) an end-organ system which can be stimulated by changes of *much less intensity* than the usual, normal end-organ system. Many possible irritants to the end-organ (eg, drugs, endocrine levels in the tissues, and chemical variations in the blood stream) as well as motion can induce subjective vertigo in such patients, when a labyrinth with normal threshold of irritability would not cause symptoms.

Electronystagmography may add information that is helpful in differential diagnosis in carefully selected, patients, but there is a tendency to overemphasize minor variations in electronystagmographic tracings, and considerable care must be taken to be sure of all the possible variables in the apparatus required.

Differential Diagnosis

Nonrotational Dizziness

It is not wise for the physician to focus all of his or her attention on hearing and labyrinthine function, because in the great majority of patients complaining of dizziness the functional examination of the ear will be normal. It is necessary to have in mind a considerable group of diseases that may cause dizziness and to know some of the more common characteristics of these diseases, so that the entire examination, beginning with the history, will narrow the field of possibilities.

The most important diseases affecting the proprioceptive system are tabes dorsalis, pellagra, and pernicious anemia. It is of value, therefore, to do a complete blood count, *serologic determination,* and neurologic examination if these diseases are suspected. Lightning pains and gastric crises often occur in tabes. Dermatitis, diarrhea, and manifestations of vitamin deficiency are usually present with pellagra. Pallor and numbness of extremities are common symptoms of any anemia. The increase in the occurrence and spread of lues in the 1980s will certainly bring, in the years to come, an increase in central nervous system disease, and the "great imitator" may once more require us to be aware of this potential cause of

dizziness.

Cerebral anoxemia is one of the most common disturbances producing dizziness. Arteriosclerosis and hypertensive cardiovascular disease often produce a more or less constant feeling of uncertainty or intermittent mild attacks of dizziness in which whirling vertigo is not found. In postural hypotension, dizziness occurs when the patient rises suddenly from a recumbent or sitting position to the erect standing position. Examination of the vascular system, including blood pressure readings, is therefore a necessary part of the evaluation of the dizzy patient. Pernicious anemia has already been mentioned in its relation to disturbance of the proprioceptive sensations. However, any anemia may produce transient or recurrent dizziness as a result of cerebral anoxemia. Again, the routine blood count may be helpful.

A group of conditions that produce sudden and slightly more prolonged cerebral anoxemia is also important. These include paroxysmal auricular fibrillation, aortic stenosis with insufficiency, attacks of arteriosclerotic heart disease (Adams-Stokes disease), and carotid sinus hypersensitivity. Because of the paroxysmal nature of these conditions, they are often confused with or misinterpreted as Ménière's disease. In some cases it may be necessary to withhold a final opinion until the patient can be observed during an attack. Fewer errors in diagnosis will be made if the physician adhere to a *rigid interpretation of vertigo* and demands specific criteria for Ménière's disease and other entities.

Paroxysmal dizziness caused by metabolic disturbances and manifesting itself through the central nervous system is less commonly seen. Migraine may produce a certain pattern that may help in the diagnosis. Hyperinsulinism from a pancreatic tumor, producing attacks of hypoglycemia, is another possibility. *Any paroxysmal dizziness that appears to have a definite relationship to hunger or occurs with a definite relationship to the ingestion of food* should at least raise this possibility in the mind of the examining physician. As in most situations in which dizziness (rather than vertigo) is a chief complaint, history is important. When hypoglycemia is suspected, a careful dietary history is essential. The usual habit of little protein for breakfast or lunch can often be easily discovered. A 5-hour dextrose tolerance test is necessary to make a certain diagnosis, and it may need to be repeated, after dietary preparation, since functional hypoglycemia is quite variable in its pattern.

Paroxysmal dizziness may be the only symptom of petit mal. Attacks of tetany from parathyroid insufficiency may also give rise to dizziness of this type. Patients with these symptoms may require long and careful study before the correct answer is found. But all the conditions mentioned should be remembered as possible causes of dizziness.

Dizziness is frequently produced by infections of the central nervous system such as meningitis, encephalitis, and syphilis. However, the other signs and symptoms coincident with these diseases will indicate the diagnosis. Dizziness is a common symptom of increased intracranial pressure, regardless of cause. *However, dizziness alone seldom leads to the diagnosis of brain tumor or other destructive disease inside the calvarium.* Other symptoms and neurologic signs are far more important in localizing intracranial disease. In these cases it may be of value to know whether there is depression of function of the inner ear on one or both sides. However, the disturbance of inner ear function in these cases does not lead to the diagnosis without more specific neurologic signs.

One of the physician's most perplexing problems is the evaluation of dizziness following head injury. It is well known that dizziness of all degrees, not infrequently lasting for months or recurring after a period of quiescence, commonly follows head injuries. When it is associated with a demonstrable, persistent hearing loss, the physician is justified in assuming that the damage is in the end-organ or the nerve. Multiple small brain hemorrhages can cause dizziness, and concussion can give rise to dizziness, tinnitus, headache, and other symptoms *without any positive physician findings.* There is no set of rules here that can aid in the evaluation, since compensation neurosis and outright malingering are common and extremely difficult to separate from true dizziness. It is best to give the patient the benefit of any doubt if positive signs of neurosis or malingering cannot be discovered.

Vertigo

Dizziness that is systematized or whirling in nature presents a more dramatic sequence of events and is seen in lesions involving the end-organ, cranial nerve VIII, and brain stem.

So much has been written and said about Ménière's disease, or labyrinthine hydrops, that any lengthy discussion here is superfluous. The syndrome is extremely common but has too often been used as a "wastebasket" into which far too many cases of paroxysmal vertigo have been dumped. If the physician adheres to the following criteria before making the definite diagnosis of labyrinthine hydrops, he or she will meet with more success in the management of this condition.

1. There must be paroxysmal attacks of whirling vertigo, usually with abrupt onset, almost always accompanied with nausea and vomiting, lasting hours, not days, and with complete freedom from vertigo between attacks.

2. There must be an accompanying perceptive hearing loss, frequently fluctuating, almost always progressive, and usually more severe in one ear.

3. There must be accompanying tinnitus, most commonly persistent between attacks and frequently fluctuating.

The associated hearing loss is often a flat, low tone perceptive loss, and the caloric reaction is typically hypoactive on both sides, although not necessarily equal. If the physician insists on these criteria, he or she may begin to fill one category in the evaluation of the dizzy patient (see Chapter 49, this vol).

Of more common occurrence is a syndrome frequently called "acute toxic labyrinthitis" or "vestibular neuronitis". In this condition the patient experiences gradually increasing whirling vertigo over a 1- to 3-day period, after which there follows a slow subsidence of symptoms. The typical and important sequence is that of steady improvement day by day after the height of vertigo has been reached. *There is no associated hearing loss or tinnitus.* This syndrome often follows an acute febrile disease but is also commonly seen after food or alcoholic indiscretion or after the use of any type of drug. Frequently the cause of the labyrinthitis is never discovered. The most effective treatment is to assure the patient of his or her impending complete recovery.

The end-organ may be suddenly and completely destroyed, either by injury or hemorrhage. The progression of symptoms here is dramatic - sudden, overwhelming vertigo, nausea, vomiting, loud tinnitus, and loss of hearing. Audiometric and caloric tests of the involved ear elicit no response, and the prognosis of hearing on the involved side is hopeless. Fortunately, slow compensation takes place over a 10- to 20-day period, and gradually the opposite labyrinth controls reasonably well the function previously governed by both labyrinths. The same symptoms may occur from sudden severance of cranial nerve VIII by basilar skull fracture. In this injury there is frequently coincidental damage to cranial nerves VI and VII because of their anatomically close relationship to cranial nerve VIII.

Tumor of the cerebellopontine angle *does not produce the severe attacks of vertigo seen in end-organ irritation.* Occasionally there may be an acute attack of vertigo caused by posterior fossa tumor when the blood supply to the labyrinth is intermittently partially occluded, but this is rare. The pressure or irritation from tumor in this area gives rise to slow, progressive hearing loss, tinnitus, and dizziness. These symptoms are not sufficient, as a rule, to warrant surgical exploration without added neurologic and roentgenologic examination. Cerebellar signs, corneal anesthesia, and increasing intracranial pressure are the common associated conditions. Cranial nerves V and VII are usually involved earliest after the cranial nerve VIII. Roentgenograms taken in the Stenvers and Towne positions are the most valuable in attempting to demonstrate erosion or enlargement of the internal auditory meatus. Special hearing testing with the Békésy audiometer may be most valuable in determining if the hearing loss has been caused by cochlear disease or is due to direct involvement of cranial nerve VIII. Pantopaque or air studies of the posterior fossa may be necessary in order to confirm small tumors.

Lesions of the brain stem are causes of whirling vertigo. Abnormality here is more likely to produce vertigo without hearing loss. The onset of vertigo in brain stem lesions may be sudden when caused by thrombosis of the posteroinferior cerebellar artery or by *multiple sclerosis.* The latter disease is not uncommon and should be suspected in persistent vertigo which does not fit the pattern of toxic labyrinthitis or Ménière's disease. A carefully recorded history, with special reference to other, suddenly occurring, neurologic symptoms that have cleared in the past, may lead the physician to suspect multiple sclerosis as the cause of the vertigo.

Finally, the possibility of dizziness as a functional symptom must be considered. Although the neurotic patient may have dizziness as one of the symptoms, it is not so common as the dizziness caused by organic disease. Functional dizziness has too often been used as a term for those cases that the physician is unable to classify. A careful evaluation, including reliable consultation, must be done before this diagnosis is made. The diagnosis should be made on positive signs of neurosis, and even then it should be made with some reservation. To label a symptom such as dizziness functional or neurotic too often dulls the investigative intelligence of the physician and leads to errors in diagnosis or neglect of other signs of organic disease.

Summary

In conclusion, a careful recording of the patient's history and an attempt to segregate whirling vertigo from other sensations described as dizziness by the patient is essential. If one

can be sure the sensation is one of true whirling or motion, the ear, cranial nerve VIII, and the central nervous system are suspect. If the sensation is *not* one of motion, the symptom may be produced by disturbances of other general body disease. The careful observer will often discover an elusive sign or symptom that has been overlooked, regardless of the thoroughness of previous examinations. The correct evaluation of the dizzy patient will be made in a higher percentage of cases if the physician adopts a broad approach and is constantly aware of the possible influence of systemic disease in the production of this symptom.