K. J. Lee: Essential Otolaryngology and Head and Neck Surgery (IIIrd Ed)

Chapter 4: The Vestibular System and its Disorders: Part I

Physiology of Vestibular System

The physiology of the vestibular system is the applied anatomy of three semicircular canals; the lateral where there is no medial, the superior where there is no inferior, and the posteriro where there is no anterior. These are at approximate right angles to each other. In addition there are two otolithic organs, the utricle and saccule.

The fundamental microscopic anatomy of hair cells of the inner ear is the same. Each hair cell has hairs or cilia of two varieties. There are stereocilia which are arranged in an ascending, pipe organ structure, and one kinocilium located at the end of the tallest stereocilia. The kinocilium is less rigid in structure and appears wavy and more flexible. It is the deflection of the cilia either toward or away from the kinocilium that stimulates the hair cell. Each hair cell has a resting electic discharge, i.e. a discharge of measurable electric activity without stimulation or deflection of the cilia. If the cilia are deflected toward the kinocilia, the rate of discharge is increased, while deflection away from the kinocilia will decrease and possibly abolish the discharge depending upon the intensity of the stimulus.

The hairs of the hair cells are inserted into a gelatinous layer which is reponsible for the manner in which the hair cells are deflected. In the semicircular canal this mass is called the cupula and extends across the endolymphatic fluid space of the ampulla to insert into the endosteal membrane. Therefore, as the endolymph flows in response to angular acceleration, i.e. rotational or caloric stimuli, rather than flowing over the cupula like a swinging door, the cupula deflects it like a sail. This endolymph flow then stimulates the semicircular ducts.

The otolithic system has, in addition to its gelatinous membrane, a mass of calcium carbonate crystals (rock pile) which renders that membrane sensitive to the effects of gravity and head tilt. The utricle is the primary otolithic structure sensitive to linear acceleration. The function of the saccule is not clear at this time. It may function similar to the utriculus and/or may function as a low-frequency sound or vibration sensor.

Since the hair cells have "polarity", i.e. where the kinocilium is located, they are lined up in functional groups. In the lateral semicircular canals the hair cells of the ampulla are all found with the kinocillia closest to the utricle. In the superior and posterior semicircular canals the kinocilia are located away from the utricle or on the crus commune side of the ampulla.

In the otolithic membranes, the hair cells are lined up with the kinocilia facing a line which almost bisects the membrane. This line is named the striola. This latter formation allows for the sensitivity seen in head tilt.

Let us examine the right lateral semicircular canal (SCC) by caloric stimulation which permits only one SCC to be stimulated. A caloric stimulus with a temperature cooler than body temperature, with the lateral SCC placed vertically causes a flow of endolymph in an ampullofugal (away from the ampulla) or utriculofugal (away from the utricle) direction. This

causes a deflection of the cilia of the hair cells of the lateral SCC away from the kinocilia. The result is a decrease or abolition of the electric discharge output of the hair cells of the lateral SCC, which in turn results in a reduction of the electric potential of the ipsilateral vestibular nuclear cells responding to hair cell stimulation. In addition, vestibular nuclear cells sampled on the contralateral side of the brain stem will show a corresponding increase in electric potential. When this potential difference occurs accross the brain stem, the medial longitudinal fasciculus (MLF) is stimulated which provides stimuli to the oculomotor nuclei, particularly III and VI, for lateral SCC stimulation.

Similar to the resting discharge of the hair cells, the extraocular muscles of the eyes have a constant group of muscle fibers contracting, and may increase or decrease the muscle mass contracting to move the eye. Thus with increased potential on the left side of the stem there is relaxation of the medial rectus of the right eye and the lateral rectus of the left eye and a corresponding increase in contraction of the lateral rectus of the right eye and medial rectus of the left eye. This results in a slow eye movement to the right. When the eyes reach a critical deviation to the side, a feedback input probably through the oculomotor system to the prepontine reticular formation evokes the rapid corrective quick component of nystagmus. By convention nystagmus is named by its quick component.

Similarly, if the other ear was stimulated with water cooler than body temperature, the nystagmus produced would be the opposite of the first ear. If the right ear was stimulated with water warmer than body temperature, then this would result in an ampullopetal (towards the ampulla) or utriculopetal (towards the utricle) movement, which results in a deflection of the cilia towards the kinocilia. This increases the discharge of the hair cell population of the lateral SCC over the resting potential. In the brain stem there is an increase in the potential of the ipsilateral vestibular nuclear cells receiving input from the SCC and a corresponding decrease in the potential of the contralateral vestibular nuclei. This reproduces the events of the brain stem similar to a cool water stimulus in the left ear. Thus the right beating nystagmus produced by cool water in the left ear will be the same nystagmus as produced by warm water in the right ear.

If water of the same temperature was irrigated into both ears, and if the response in both sets of hair cells was equal, the resultant effect in the brain stem vestibular nuclei would be no change, therefore no stimulation of the MLF and no nystagmus produced.

Similarly, a lesion in the inner ear, vestibular nerve, vestibular nucleus, or internuncial nerve tracts of the vestibular nuclei can produce a differential across the brain stem resulting in a spontaneous nystagmus. If the differential were not present except under stimulus conditions then this may be evident by a reduction in caloric response, or nystagmus on the changing of the head position.

Thus the effects of caloric stimulation can be understood as well as the rotary stimulation of the lateral SCCs. The posterior and superior SCC also can be stimulated but produce an oculomotor response which at present is difficult to sort out for clinical diagnostic purposes.

The otolithic system produces ocular counterrolling which allows for some measure of function. This technique is cumbersome, difficult, and can be fraught with interference by other oculomotor phenomena.

Methods of Testing the Vestibular System

At this time any test of the vestibular system without an electrical recording of nystagmus behind closed eyes may mislead the clinician in the proper management of the patient. Electronystagmography (ENG) is an electronic method of producing a permanent record of eye movement. This movement can occur with or without the eyes open, in any position of the head, and under any circumstance of stimulation of the vestibular system. A method of vestibular evaluation is described along with the derived conclusions of the test.

The patient is ushered into a room that is permanently set up as a clinical vestibular laboratory. The technician or physician explains all aspects of the procedure as it progresses, both to relieve apprehension and to act as a form of constant alerting for the patient. The patient is seated on the ENG table and the ears are examined for obstructive cerumen, tympanic membrane perforations, or the presence of mastoid cavities. After that, the patient lies supine on the table and the skin is cleansed and dried where the electrodes are to be placed. The electrodes are applied to the skin after filling the electrode cup with a coupling electrode jelly or paste. The ground electrode is placed above the bridge of the nose. The electrodes for recording horizontal nystagmus are placed on the skin as close as possible to the outer canthus of both eyes. The electrodes for recording vertical nystagmus are placed above and below one eye. If necessary, and if the recorder has the capability, each eye motion can be separately recorded with an array of electrodes around the eye. Once the electrodes are applied, the gain of the recorder is adjusted to produce a specific deflection of the pen on the paper in keeping with the magnitude of a calibraing eye movement. This is accomplished by following alternating lights a known distance apart. Once calibrated, the eye movement can be recorded behind closed eyes. The electric potential change being recorded emanates from a potential between the cornea and retina moving within the recording field.

Once calibrated, the corneoretinal potential may vary with the amount of light stimulation of the eyes. It may be necessary to alter the gain from time to time during the testing time in a recalibration procedure.

The patient can then be placed in various positions to see if nystagmus can be recorded. The nystagmus may occur in only the supine position, the right lateral, the left lateral, or all of the position. It may be beating all in the same direction, or the direction of beating may change in one or more positions.

The velocity of the nystagmus may change significantly from one position to the other. Turning the head (neck torsion) also may produce nystagmus, turn off the nystagmus, or enhance any nystagmus already present by just lying on that side. The presence of nystagmus in any of the foregoing is abnormal. The significance, however, is not clear. Some form of spontaneous positional, or positioning nystagmus may be present and is abnormal. It may not represent a serious problem, or it may be the only clue to substantiating an organic vestibular disorder and uncovering a potentially serious health problem, such as an acoustic tumor.

The caloric tests have been recognized as the most beneficial in vestibular diagnosis. Each ear can be stimulated separately. By using a bithermal stimulus (30°C and 40°C) the same ear can be stimulated and produce an opposite direction of beating of nystagmus. Thus with the patient's head elevated 30°C from the supine position, the lateral SCC is positioned in the vertical plane, and the maximum effects of stimulation of the endolymph can occur. A unilateral weakness of an ear is recognized when there is less nystagmus produced with both the warm and cool stimulation of one ear compared with the other ear. At times the response is not clear and a mathematical formula comparing the difference in velocity of response from one ear to the other, expressed as a percentage of the total response, may shed light on the quantity of difference. Thirty percent was derived in some studies while others use a 25 or 20% difference to recognize a reduced vestibular response (RVR).

At times there is more nystagmus produced in one direction than the other even though the difference between ears calculates as normal. The tendency to have a significantly greater amount of right-beating nystagmus, for example, is named a "directional preponderance" (DP). Where it is not obvious, a formula also may be used expressing the difference in right-beating nystagmus from left-beating nystagmus as a percentage of the total response of all four bithermal stimuli. Some investigators use from a 30% down to a 15% difference as a significance level. When present, and pathologic or significant, it means that the system is not functioning normally, but it has no localizing value as to right or left, central or peripheral.

Clinical Presentation

History

1. With problems of dysequilibrium, the ability to give clearcut historical information is dependent upon the patient's vocabulary and previous experience of vestibular stimulation. Some may express being "just dizzy"; others may feel a rotary experience. With others there is an illusion of movement of the environment without rotation. Unsteadiness, light-headedness, walking on a cloud, or a floating feeling are other expressions of symptoms which are often referable to abnormalities in the vestibular system.

2. Hearing loss, tinnitus or fullness of the ear(s) will often lead to an otologic origin.

3. Difficulty with speaking or swallowing while walking, otherwise exercising, or expanding cardiac output may be a sign of vascular insufficiency.

4. The character of the dysequilibrium such as onset, duration, frequency, disability, precipitating circumstances, aggravating circumstances, and those which improve the symptoms may provide clues to the cause.

5. The family history may provide information about neurofibromatosis, diabetes, otospongiosis, or other factors.

6. Surgical or accidental trauma, as well as the use of drugs for extended periods, also may provide etiologic clues. Associated medical illnesses should be catologued according to factors in the symptoms.

Examination

1. The ears are examined for evidence of trauma to the tympanic membrane, serous otitis media, or evidence of chronic otitis media. The sensation of the posterosuperior aspect of the skin of the medial portion of the external canal may be examined for sensitivity.

2. The use of tuning forks will give clues to conductive or unilateral sensorineural hearing loss. The response of the ears to the forks may outline the anticipated audiometric findings.

3. Next, the remainder of the cranial nerves are examined. The olfactory nerve is difficult to fully examine, but the third, fourth, and sixth are easily examined in searching for gaze nystagmus or any other eyes-open nystagmus. Cranial nerve II is judged by confrontation. The fifth cranial nerv, so important in acoustic tumor diagnosis, can be assessed easily in three areas: corenal sensitivity, light touch, and pinprick in all three divisions. The facial nerve may not display evidence of paralysis but a difference in the width of the orbital fissures or a flattening of the nasolabial fold may provide subtle findings of its early compromise. Nerves IX, X, XI, and XII all become part of the remainder of the head and neck examination.

Audiology

1. Pure tone (air or bone), along with speech reception and discrimination, provide the bare bones and minimum of the auditory function evaluation.

2. Impedance testing including tympanometry and stapedial reflexes, both ipsilateral and contralateral are also essential. In addition, where the stapedial reflexes are present, decay also should be sought.

3. Auditory brain stem evoked potentials will add further knowledge toward understanding the neurophysiology and neuropathology of the hearing mechanism.

4. There are a host of other tests such as SISI, Bekesy's, ABLB, difference limen testing, etc, which have been by and large supplanted by the more modern previously described tests.

Electronystagmography (ENG)

1. The first part of the test occurs with calibration. Consistent overshoots, if present on calibration of horizontal eye movements with small enough calibrating lights, are a sign of ocular dysmetria. This should be considered a sign of a central lesion.

2. Once calibrated, a search is made for nystagmus with the eyes closed in the supine and left and right lateral positions. A portion of the tracing will also include eyes open to see the effect of ocular fixation. This also tends to reduce the intensity of corneoretinal potential change with prolonged darkness. Nystagmus, when found in these positions, is abnormal and this is the most important contribution. When it occurs only once with one ear down, this may provide an additional clue along with the history, clinical examination, audiology, and caloric

response to the origin of the dysequilibrium. At times nystagmus will be found in all position. When it is the same direction and similar velocity, then this can be named spontaneous nystagmus. Spontaneous nystagmus is abnormal when present. It has no localizing value as to the site (central or peripheral) or the side (right or left) no matter which direction the nystagmus.

When the nystagmus is the same direction in all positions but varies in velocity, it is a direction-fixed nystagmus. The majority of such findings are peripheral, but not always. At times when there is a large difference with one ear down compared with the remainder of the positions, the difference then may have diagnostic value. The ear down is not always an indication of the pathologic site. This must be correlated with other findings before arriving at a conclusion. To arrive at a decision based upon the position response alone will be erroneous in a small but significant number of patients.

When the nystagmus is present in all positions but changes in direction, it is named a direction-changing positional nystagmus. While this is thought to be a central sign, it is sufficiently inaccurate to be relied upon as a central sign. Of importance is the right-beating nystagmus with the left ear down only, and the left-beating nystagmus with the right ear down only. This is suggestive of the second phase of positional alcohol diffusion, becoming gravity dependent and representing a peripheral abnormality of ingested alcohol over the prior 24-48 hours. Careful questioning is important as to ingestion of substances which could alter the vestibular evaluation.

3. While the lateral positions give evidence mainly about the effect of the otoliths, the addition of turning the neck into these positions adds input from the cervical nerves. These can either produce a nystagmus not previously present, enhance or reduce a previously existent nystagmus, or turn off or produce a nystagmus in the opposite direction. This effect is very potent and must be considered in the development of the symptom complex. The possible effect upon the vertebral arteries also should be considered. The latter tend to play a lesser role in the production of symptoms as a whole.

Alternate Binaural Bithermal Calorics

The caloric stimulus provides the most information about the functioning of the vestibular system. Two temperatures of water, one above and one below body temperature, must be used. Since the response produced will be nystagmus in one direction with one temperature and the other direction with the other temperature, two facets can be examined. Both the warm and cool responses may be reduced in one ear compared with the other. In some instances the use of Jongkees' formula may provide a numerical percentage difference not readily apparent from "eyeballing" the nystagmus. Where there is a directional preponderance than the cool in one ear and the warm in the opposite produces a nystagmus response in which the intensity is greater than the opposite-beating nystagmus. This points out fallacy of using a screening warm or screening cool stimulus. If one were to examine a single screening technique, then 60% of the time the warm hypoactive response would point to the lesion. This leaves an accuracy of about 40% if the cool stimulus were used as a screening temperature. Some laboratories, if the cool responses were low enough, would add an ice water stimulus to determine if more function could be found.

Simultaneous Binaural Bithermal

By stimulating both ears simultaneously with the same temperature of water (i.e. 30°C) and looking for nystagmus followed, after an appropriate rest period, by a simultaneous stimulation of the other temperature of water (i.e. 44°C), a more sensitive stimulus was found. The nystagmus responses are divided into four groups, or types, on the basis of the presence of nystagmus and its direction. By stimulating both ears simultaneously, the brain stem receives equal and opposite stimuli. The vestibular nuclear cells perform the task of processing the input. If it is entirely equal and opposite, then no stimulation of the MLF occurs. If the response in the brain stem results in a difference, then the MLF is stimulated. As such, the results of the stimulus produce diagnostic information 55% more sensitive than the alternate binaural bithermal method alone. However, if the simultaneous stimulus were used alone, then an error of at least 2.5% would be made. So, the simultaneous binaural bithermal stimulus is recommended as a 6.5 minute addition to the alternate stimulus in the course of vestibular evaluation.

Massive Caloric

Some patients who have had previous attempts at labyrinthectomy, or with apparent absent function on bithermal testing, may require further testing. If the dizziness persists and is disabling, a more thorough labyrinthectomy or vestibular nerve section may be considered. A search is made for any residual function to be destroyed. A massive caloric stimulus may demonstrate this function. Two temperatures of water are used; ice water and 46-48°C. If there is a preexisting nystagmus, the the first temperature of water delivered to the ear in question should increase the velocity of the preexisting nystagmus. This is followed immediately with the other stimulus. The recorder remains on during the entire stimulus time. The volume delivered is about 300 mL in about 30 seconds for each stimulus. A search is made for the production of and reversal of nystagmus as a demonstration of residual caloric function. As a footnote, the integrity of only the lateral SCC is examined and if the massive test fails to stimulate any residual function, any further decision making will require information from other areas.

Dysrhythmia

There are two types: alerting and central. The alerting type is normal and occurs with a bored or poorly alerted patient. The recording shows bursts of clear, regular, sawtooth nystagmus interrupted by intervals of no nystagmus at the point in the response where the nystagmus should be brisk. This can be alleviated by changing the alerting task or making it more difficult.

Central dysrhythmia is recognized by nystagmus beats whose amplitude, velocity, and morphology bear no resemblance to each other. That is, the central regulatory mechanism is unable to produce even and regular nystagmus beats.

Ocular Fixation Suppression

When the nystagmus produced is at its most brisk, then opening the eyes should reduce or eliminate the nystagmus by mere ocular fixation or the attempt at fixation. This finding occurs with a normal vestibular system or in one with a peripheral vestibular disorder. In some central lesions the caloric-induced nystagmus will not appear until the eyes are opened, will significantly increase when the eyes are opened, or will show no signs of suppression of the already present, induced nystagmus. The test should be performed on all four caloric responses to the alternate binaural bithermal stimulus. When present this is a reliable sign of a central disorder.

Future Tests

With the explosion of the computer field and electronic and mechanical technology, newer tests of the vestibular system will become available. At this time (1981) the use of rotation chairs with a computer appears to provide additional helpful clinical information. While both ears are simultaneously stimulated, the information on the laterality of a lesion cannot be accurately deduced. However, by looking at the gain of the vestibulo-ocular reflex with differing frequencies of rotary stimulation, the responses can be classified into normal, central, or peripheral in fairly clear-cut categories. By looking to the degree of labyrinthine preponderance, the degree of compensation of a vestibular lesion then can be assessed. In many instances a patient may demonstrate a stable abnormality in the ENG and yet be fully compensated and symptom free, or fully symptomatic and disabled. The abnormality on the ENG may be exactly the same. The degree of labyrinthine preponderance has shown evidence of compensation when it is present.

Tracking tests using a pendulum or optokinetic stimulus have demonstrated little or no usefulness in clinical testing. The use of a computer generated, LED screen stimulus and a computer analyzed response may provide more accurate clinical data. Unfortunately, as with all-eyes-open voluntary tracking tests, patient cooperation significantly factors into the validity of the test results.

Clinical Entities

Metabolic Vertigo

There are no clinical symptoms which separate metabolic vertigo from other forms. A prerequisite may be an abnormally functioning vestibular system. In this instance the metabolic factor exaggerates of interferes with the compensatory mechanisms and brings about the symptoms. It is imperative to include metabolic studies. These may indicate a contribution to the symptoms that when eliminated may improve the symptoms to a livable rather than disabling level. Elevated blood fats or a fluctuating blood sugar commonly can be found where there is a diet too high in carbohydrates and/or animal fats. Dietary modification often will result in a striking improvement in symptoms.

Hypothyroidism is an extremely rare but definite cause. Many times the patients are not otherwise clinically hypothyroid. The inclusion of a test for hypothyroidism probably is justified.

Lastly, allergic causalities are very elusive in the management of a dizzy patient. A screeing IgE may give a clue; RAST or skin testing may provide more precise findings about an allergic cause and its treatment. In the absence of a clear-cut history, the allergy evaluation is indicated in the absence of any other clearly defined cause.

Perilymph Fistula

This finding as a cause for vertigo in the absence of hearing loss is found more frequently than expected. The history should be straightforward for trauma or air travel and the resultant symptoms clearly follow. Such is not always the case since a sneeze or vigorous blowing of the nose may be the inciting event. The resultant vertigo may not occur for some period.

The clue in the history is one of an episodic nature usually related to exertion. Many patients will be asymptomatic on awakening in the morning only to have symptoms appear once they are up and around. A positive fistula sign with or without ENG is helpful, although a negative sign does not rule out a fistula.

Associated symptoms of ear fullness, tinnitus, and mild or fluctuating hearing loss will help to localize the ear. In addition, the caloric findings, especially the simultaneous calorics, are most helpful. Many patients will demonstrate nystagmus with the affected ear down. However, this alone is not a reliable sign to localize the pathologic ear.

Ménière's Disease

The histopathologic temporal bone finding correlated with this disease is endolymphatic hydrops. In its pure form the hydrops should have no traceable cause such as suppuration, trauma, otospongiosis, and syphilis.

The symptoms, where complete and classically present, include fluctuating sensorineural hearing loss, fluctuating tinnitus, and fluctuating fullness in the affected ear. In addition, as the tinnitus, fullness, and hearing loss intensify an attack of vertigo will follow. These tend to occur in episodes. The process may spontaneously remit never to occur again and leave no residual or perhaps a mild hearing loss and tinnitus. It also may go on with recurring disabling vertigo and progressive hearing loss.

The idiopathic form is not commonly bilateral. If bilateral, a causality needs to be conscientiously ruled out.

The symptoms also may occur alone as in cochlear Ménière's disease; vestibular Ménière's disease, or Lermoyez's syndrome. In general, these will have ear fullness as the common denominator.

The audiology findings, where present, will show an early fluctuating low-tone sensorineural hearing loss. The discrimination initially is well preserved. Site-of-lesion tests will demonstrate a cochlear lesion. With serial hearing tests and no remission, the hearing loss will fluctuate with changes in the pure tones, or in the speech discrimination scores or both. Eventually the hearing levels will decline, with mainly evidence of fluctuating discrimination

scores. Untreated, the discrimination scores eventually will yield no useful hearing ability even with amplification.

The ENG findings commonly show very little between the initial episodes. During the attack there may be active spontaneous nystagmus with direction changing components even in the midst of caloric testing. Alternate bithermal calorics may reveal an abnormality about 50% of the time, while simultaneous bithermal calorics will increase the yield to 80%.

The natural history is of a final and complete remission in about 60% of the patients. The end point in the hearing loss and vestibular disorder varies with the point of the remission. Residual tinnitus also may be a disturbing factor.

Since the stage at which a spontaneous remission will occur cannot be predicted, several medical and surgical therapies have evolved to alter the end results. Each therapy has its strong advocates. The presentation of the disease with its long remissions and exacerbations, and the inability to measure the degree of hearing loss or vestibular disturbance on line, make it an enigma for therapy.

The medical therapies are aimed at the symptoms and include vestibular suppressants, vasodilators, and diuretics.

The surgical therapies are either destructive, conservative, or preservative. The first includes labyrinthectomy or translabyrinthine eight nerve section where there is no useful hearing. Selective destructive procedures where there is useful hearing include midfossa vestibular nerve section, ultrasonic irradiation of the labyrinth, or cryosurgery.

Conservative procedures include those performed on the endolymphatic sac. These range from sac decompression to endolymphatic-mastoid shunts to endolymphatic-subarachnoid shunts. The endolymphatic-subarachnoid shunt appears to be directed at a correction of the resultant mechanical changes seen in the histopathology of the hydrops.

The tack procedure appears to result in a steady decline in the hearing, while there has been little experience with cochleostomy as a treatment.

As statistical methods and means of evaluation improve, the results of the various therapies will be clarified and the total management of this enigma standardized.

Acoustic Neurinoma

This benign tumor has its origin most commonly in the superior vestibular division of the eight cranial nerve. Because these are slow growing, very few symptoms arise as this division of the eight nerve is being compromised. Compensation for the slow loss of the nerve results in no marked vestibular complaints. As the enlarging tumor spills over into the cochlear division of the eight nerve or compromises the artery to the inner ear, hearing symptoms will manifest themselves. This will produce either unilateral tinnitus or hearing loss or both. Occasionally the findings may be indistinguishable from a hydrops. With time there is a progressive hearing loss with discrimination affected long before a total hearing loss occurs.

As the tumor enlarges it compresses the surrounding nerves. The seventh (facial) nerve, primarily a motor nerve, seems to be resistant to the pressure effects of an enlarging tumor. Even though this is the nerve next affected, visible signs of facial nerve palsy are very late. More commonly, the effect of pressure on the fifth (trigeminal) nerve is demonstrated by altered corneal sensation, the first modality of this nerve that is usually affected. Later there may be symptoms of numbness in any or all divisions of the nerve.

The next area affected varies with the size and direction of tumor growth. If the growth is more medial, the obstruction of flow of the cerebrospinal fluid may result in hydrocephalus; if more posterior, then cerebellar signs may be elicited. An inferior growing tumor may give rise to findings in cranial nerves IX, X, XI, and XII. At the extreme of the very large tumor there may be signs of the effect on all of the above nerves.

These tumors also may arise from the cochlear nerve where symptoms will appear clearly, or the inferior vestibular nerve where the findings are characteristically very late.

The early diagnosis of these tumors requires a high index of suspicion and diligence in the diagnostic pursuit. When the tumors are quite large they represent no diagnostic dilemma.

The audiologic evaluation may vary from normal hearing in a normal pure tone audiogram with poor speech discimination, to a pure tone sensorineural hearing loss and poor or no speech discrimination. A search for stapedial reflexes with the impedance bridge may show reflexes present at normal levels without evidence of decay in about 18% of the tumors. The reflexes are helpful when absent or show evidence of decay of the reflex when the behavioral pure tones are in the normal range.

Auditory brain stem evoked potentials also may increase the index of suspicion. This test although very valuable is known to have its false-positive results, but more importantly, its false-negative.

The vestibular evaluation heightens one's suspicions when there is an absent caloric response in the suspect ear in the absence of any history of dysequilibrium. Alternate binaural bithermal testing observes the function of the superior vestibular nerve while the simultaneous stimulus uncovers more abnormalities consistent with the tumor.

The radiologic evaluation provides more reason for suspicion and finally the indication for surgery. Plain roentgenograms and more accurately complex motion tomography of the temporal bone may show an enlarged internal auditory canal. While a 1.5-2.0 mm difference is accepted as the radiologic normal variation, if this occurs on the suspect ear, the coincidence is too great to overlook. Large differences also may be present.

With the suspicion present, CT scanning with intravenous contrast is the first step. This will identify tumors of 1.5-2.0 cm or larger. To identify smaller tumors some contrasting substance needs to be physically introduced into the angle. Currently, air or CO_2 with high-resolution CT scanners has outlined smaller tumors or those within the internal auditory canal. False-positive findings have been found as more experience is gained with this modality.

Positive contrast iophendylate in the posterior fossa with plain or complex motion tomography also will outline a mass in the canal or angle. This method also uncovers vascular loops and arachnoid cysts which cannot be differentiated from the tumor until the time of surgery.

The surgical treatment has advocates among the neurosurgeons and the neurotologic surgeons. The area of agreement in the surgical treatment demands the use of the operating microscope. The patient in a sitting position, as used by some neurosurgeons, may result in problems of quadriplegia or air emboli. Thus the supine or prone position may be preferred.

When there is hearing worth preserving and the tumor is within the internal auditory canal, the midfossa approach has the best chance for tumor removal and preservation of the hearing. When the hearing is good and the tumor is well into the cerebellopontine angle there are advocates for suboccipital removal. The exact number of patients whose normal hearing has been preserved has not been tabulated. In addition, the mobidity may be increased.

The translabyrinthine approach can be used when there is no useful hearing. Some surgeons will recommend the translabyrinthine approach because they experience less morbidity and find it impossible to preserve hearing when the tumor is out of the internal canal. Others may recommend the suboccipital approach.

Some combine the advantages of the translabyrinthine and suboccipital approaches into a one-stage removal of very large tumors.

Multiple Sclerosis

This neurologic disease with lesions disseminated in space in the neurologic system in time may present with vestibular symptoms.

The lesions may be exaggerated by warming the body temperature. The electronystagmography may show anything from normal findings, to peripheral findings, to central findings. Auditory brain stem-evoked potentials may show delay of central conduction. More likely, there is significant delay of the visually evoked potentials.

Evidence of anterior internuclear ophthalmoplegia on testing eye movements also may give a clue to this neurologic disease.

Syphilis

The otologic findings generally consist of bilateral clinical endolymphatic hydrops. There is significant hearing loss and usually bilateral absent caloric function. The patients as a rule are in the mid-40s. The serologic tests which rely on the presence of reagin are usually negative. The specific treponemal tests such as the fluorescent treponemal antibody absorption or hemagglutination inhibition are positive.

The treatment consists of a course of penillin therapy and desensitization to this drug, if possible, in those patients allergic to penicillin. The use of steroids may result in a dramatic

improvement in hearing and reduction of vestibular symptoms. Usually the steroids must be maintained indefinitely to retain the clinical improvement.

Posttraumatic Vertigo

This comprises a history of head trauma followed by a number of possible symptoms. The common denominator is dysequilibrium. If there is a total hearing loss then the use of vestibular suppressants may result in a cure which is sustained after cessation of the suppressants. In some instances where there is no cure, a labyrinthectomy or eighth nerve section will ameliorate the symptoms.

Occasionally there is a progressive hearing loss which therapeutically is managed in the same manner as the immediate hearing loss and vertigo.

Delayed endolymphatic hydrops, which is resistant to medical therapy, may develop. In this instance endolymphatic sac surgery will improve the symptoms, provided there is no fracture displacement through the endolymphatic duct. This latter findings may not be evident on a precise radiologic study or at the time of surgery.

The dislodging of otoconia which roll toward the ampulated end of the posterior semicircular canal has been postulated as a cause for positional vertigo of a posttraumatic type. The nystagmus is said to occur with the affected ear down, and singular nerve neurectomy is recommended as the therapy. While this may occur there are some errors in taking the undermost ear as the pathologic site. In addition, the use of habituation to reduce the symptoms may supplant surgery as the mode of therapy.

Otospongiosis (Otosclerosis)

There appear to be three areas where otospongiosis may bear a relationship to dysequilibrium. The first occurs in relationship to the fixed footplate. By having a fixed footplate, there may be a change in the fluid dynamics of the inner ear giving rise to vestibular symptoms. In a highly reliable number of patients the symptoms are cleared by stapedectomy.

Sometimes vertigo may begin after stapedectomy. This may occur with a perilymph fistula which requires revision and repair. A total, irreversible loss of hearing with vertigo also may occur, and a destructive procedure of labyrinthectomy or eighth nerve section is indicated if the vestibular suppressants fail to control the dysequilibrium.

Endolymphatic hydrops secondary to an otospongiotic process has been described from histologic evaluation of the temporal bone. This must be considered in the etiology of hydrops and may respond to fluoride therapy.

The coexistence of otospongiotic foci around the vestibular labyrinth with elevated blood fats or abnormalities of blood sugar may give rise to vestibular symptoms. The effective treatment here requires fluoride therapy and attention to diet.

Vascular Insufficiency

Vascular insufficiency also may give rise to vestibular symptoms. This usually occurs in the elderly patient or those with other evidence of extensive vascular disease. The vestibular findings may show bilateral absent caloric function. Angiography in the absence of focal neurological findings is probably not justified nor likely to lead to a different therapeutic regimen.

The subclavian steal syndrome may be found by differential blood pressure in the arms or direction Doppler confirmed by specific aortic arch angiography and surgical treatment.

In some instances, anticoagulation may be tried as a diagnostic test and continued if therapeutically indicated.

Vestibular Neuronitis

This clinical entity begins with a nonspecific viral illness followed in a variable period of up to 6 weeks by a feeling of dysequilibrium which may be disabling. There are no symptoms or findings in the cochlear system. Electronystagmography demonstrates a unilaterally reduced caloric respone. The remainder of the evaluation is negative for a cause. In most patients, the symptoms, in relation to vestibular compensation, clear with time. This may be hastened by the use of effective vestibular suppression for a period of up to 6 weeks.

A small percentage of patients so afflicted will not respond to vestibular suppression or to vestibular compensation. In these, after an appropriate observation period, a midfossa vestibular nerve section is indicated. Abnormal myelination has been found in some of these nerve specimens.

Otitis Media

Either suppurative or serous otitis media may have vestibular symptoms in association. In serous otitis media the presence of fluid in the middle ear restricting the round window membrane, and/or serous labyrinthitis from diapedesis of protein through the round window membrane, may be responsible for the vestibular symptoms. Removing the serous fluid either medically or surgically gives rise to a remission of the dizziness.

In the presence of suppuration there may be a reversible serous labyrinthitis, or an irreversible suppurative labyrinthitis and the more extensive sequestrum, with a dead ear and facial nerve palsy. In these instances judgment of the disease and its effects will determine the proper treatment.

Ototoxic Drugs

These predominantly aminoglycoside antibiotics are usually used in lifesaving situations where no other antibiotics are judged to be as effective. Vestibular evaluation utilizing ENG will demonstrate no response to warm, cool, or ice-water testing. Minimal response may occur in some instances. The patients may range from being totally incapacitated to being only marginally disabled. The main symptom is oscillopsia, and results

from lack of otolithic input to allow the eyes to maintain a level horizon while the head is bobbing up and down as the individual walks. Sometimes the usual vestibular suppressants may aid the patient. In other instances one is frustrated by an inability to adequately treat this condition.

Drugs

A complete drug history is essential as many drugs alone or in combination may act centrally or peripherally to induce symptoms of dysequilibrium.

Alcohol is a common drug whose effect is clearly on the end organ in the second phase of positional alcohol nystagmus. This shows a left-beating nystagmus while lying on the right side and the opposite while lying on the left side. There is no nystagmus in the other positions. This effect can be neutralized by ingestion of heavy water before alcohol ingestion.

Cervical Vertigo

An injury to the neck affecting the spinovestibular input can give rise to dysequilibrium. As more of these patients are studied, another lesion somewhere in the vestibular system also is being found. However, the neck lesion brings out the dysequilibrium and if adequately treated can relieve the symptoms.

Vertigo in Epilepsy

Dysequilibrium as a symptom of epilepsy is seen in two forms. The first is an aura of a major jacksonian seizure. The second is the momentary, almost petit mal, seizure whose entire brief movement is experienced as dysequilibrium. The diagnosis of this latter form may require a sleep EEG. These patients will respond to usual seizure control therapy.

Cardiovascular Causes

Arrhythmias usually may produce dysequilibrium. These rarely presents to the otologist but are seen in consultation with the cardiologist. However, consideration must be given when seeing a new patient with dysequilibrium.

Intracranial Tumors

There is a small but definite number of intracranial tumors which have hitherto gone undiagnosed. These have some degree of dysequilibrium associated with them. The use of CT scanning, without and with intravenous contrast in selected patients, may help to identify these otherwise silent lesions. The type III response to simultaneous binaural bithermal stimulus may be helpful here.