NEURO-OTOLOGICAL EXAMINATION

with Special Reference to Equilibrium Function Tests

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Foreword

Twenty years ago vestibular function tests were the only standard examination for patients complaining of vertigo. Recently, however, it has been generally agreed that vestibular tests should be only a portion of the examination related to equilibrium function. Since vertigo or dizziness is one of the subjective symptoms of vestibular dysfunction it seems most important to obtain some objective correlates of this complaint. Many different methods of analysis have therefore been adopted for qualitatively and quantitatively evaluating equilibrium dysfunction. Not only the location of the underlying pathology but also the nature of the lesion must be determined in order to effect a correct diagnosis and treatment. It is thus apparent that traditional vestibular tests do not suffice: improvement in the currently available examinations and development of new techniques are strongly needed. Further, the results of such functional analyses including audiologic tests must be interpreted on the basis of a systemic neurological understanding. The term "neuro-otology" is based on this concept.

The authors of this book are all actively engaged in research in this newly expanding field of neuro-otology and their original contributions in this area have been quite eminent. The current status of our understanding of neuro-otology is delineated in this volume and a prospectus for future research is also noted.

It is my pleasure to recommend this epoch-making volume to specialists all over the world. I am convinced that this publication will contribute significantly to the development of neuro-otology.

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Foreword

The vestibular system includes a complex of central connections which form various pathways and reflexes influencing three major systems; the oculomotor, the somatic motor, and the autonomic nervous systems. The cerebellum and the cerebrum have also been shown to be connected with the vestibular system. The conventional vestibular function tests previously employed, however, have been centered around investigating the peripheral vestibular organs, while the relationship of these organs to the brain as an entity has been rather neglected.

During the past twenty years, the fundamental significance of vestibular function tests has been re-evaluated and this has given rise to several new methods of testing. The development of these modern methods has been supported by the progress in our understanding of the anatomy, physiology and pharmacology of the brain. Internists, neurologists and neurosurgeons have also contributed to this development. Modern equilibrium function tests endeavor to assess the equilibrium state of the whole human body without reference to a particular set of organs and should not be bound by the framework of a particular medical subspeciality. The scope of equilibrium function tests has thus been greatly enlarged and this is why the title of this monograph "neuro-otology" was chosen in preference to the title "vestibular function tests".

This book aims at clarifying the significance of each equilibrium function test. The clinical interpretation of the test results is provided and the step-by-step explanation of procedures for important test methods are explained. The rich supply of clinical material covered in the book will also assist in gaining an understanding of the clinical application of the tests.

I fully expect to see this book take its place as a classic in the field of neuro-otology.

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Preface

In 1968 the "Neurotological Examination with Special Reference to Vestibular Function Tests" was published by Drs. T. UEMURA, J. SUZUKI and J. HOZAWA in Japan. It was based on the contents of the symposium on the "Problems in utilizing equilibrium function tests as routine tests" held at the 67th Annual Scientific Meeting of the Japan Oto-Rhino-Laryngological Society on May 5, 1966. The book received a wide welcome not only by otologists but also by neurologists, neurosurgeons and others. This encouraged us to promote publication of the English edition.

A considerable portion of the book has been rewritten and the chapter on the neurologic examination by Dr. S. M. HIGHSTEIN has been added as this examination is indispensable for obtaining the correct diagnosis in patients with vertigo and/or disequilibrium. The standardization of the neuro-otological examination has, however, not been completed. Parts of the examination are still being performed by individual physicians according to their personal experience and preference, and are not necessarily based on current knowledge of vestibular mechanisms. It is our hope that this book will stimulate discussion on many of these individual viewpoints and will contribute to the synthesis of a standard method for the neuro-otological examination.

We wish to express our sincere thanks to many friends and colleagues, without whose cooperation this book could not have been completed. We are particularly grateful to Drs. M. MORIMOTO and I. KIRIKAE for their contribution of forewords, and also to Dr. B. COHEN, Professor of Neurology at the Mount Sinai School of Medicine of The City University of New York, for his valuable suggestions and advice. The authors would like to express their gratitude to Ms. R. SEKHAR for editorial assistance. Gratitude is also due to the cooperation and assistance of the staff of the Igaku Shoin Ltd.

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Introduction

Selection of the Test Methods in the Neuro-Otological Examination

The neuro-otological examination is performed for the purpose of diagnosis and to evaluate the course of treatment. As it is usually necessary to compare the results of repeated examinations, a small number of carefully selected tests should be employed in order to efficiently and accurately obtain the optimum amount of information.

Before any special tests are employed a routine medical history and physical examination should be performed. The examiner then evaluates the patient's symptoms with reference to the vestibular system if indicated and tries to reach a tentative diagnosis. Further appropriate diagnostic tests are then employed to determine the location, extent, and etiology of the pathologic process.

The following three conditions concerning the location of the process should be satisfied.

1) differentiation between vestibular and non-vestibular disorders
2) differentiation between peripheral and central vestibular disorders
3) differentiation between the affected and normal sides.

1), 2) and 3) can usually be accomplished by performing a series of clinical examinations including a basic neurologic examination. Next, an attempt should be made to evaluate the extent and etiology of the lesion. Laboratory tests are often helpful in this regard. An example of the procedure is listed in Table 1.

Indications for the Neuro-Otological Examination

All diseases connected with vertigo and dizziness*, equilibrium disturbance, tinnitus and deafness may be referred to the otologist and *should be studied by the neuro-otological examination.*

VERTIGO AND DIZZINESS OF VESTIBULAR AND NON-VESTIBULAR ORIGIN

When the origin of an equilibrium disturbance is in the vestibular system, it is called a vestibular equilibrium disturbance, and is differentiated from a non-vestibular disturbance. Dizziness of ocular origin or psychogenic dizziness, usually continuous rather than episodic, belongs to the latter category of illness. The neuro-otological examination is useful in differentiating a vestibular disturbance from a non-vestibular disturbance i.e. positive results indicating vestibular dysfunction may be taken as synonymous with vestibular dis-

* The words vertigo and dizziness are often used interchangeably; however vertigo implies a sensation of turning which is not necessarily present in dizziness. Neither word has any etiologic implications.

Table 1 An approach to the patient. When a patient comes to the doctor with complaints of vertigo, tinnitus and hearing loss, the following procedures are performed in order to determine the cause and extent of the illness, and to follow the effects of any treatment which is necessary.

Patient

1. *History-taking*
2. *General physical examination*
3. *Diagnostic tests*

a. Ascertain the location of the illness (Differential diagnosis, peripheral or central?).

Appropriate tests:
Hearing test,
Spontaneous-, positional-, and positioning nystagmus test,
Self-recording cupulometry,
Caloric test,
Optokinetic nystagmus test
Neurologic examination

b. Ascertain the cause and extent of the illness

Appropriate tests:
Test for focal infection (oral cavity, teeth, paranasal sinuses and tonsils).
Otoscopy and X-ray examination of temporal bone.
Laboratory examination (Autonomic nervous function).
More detailed investigation of results of equilibrium and neurologic examinations.
Angiography or encephalography in cases with suspected endocranial lesion.

Diagnosis and determination of *course* of treatment. For example, the diagnosis of Ménière's disease is established.

4. *Serial tests* conducted after the beginning of treatment(Evaluation of effects of treatment and estimation of prognosis).

Appropriate tests:
Spontaneous-, positional- and positioning tests,
Rotation test,
Optokinetic nystagmus test and eye-tracking test, hearing test.

turbance. If the results of all neuro-otological examinations are equivocal, a psychogenic or other disease process may be suspected.

PERIPHERAL AND CENTRAL VESTIBULAR DISTURBANCES

When the origin of an equilibrium disorder is in the labyrinth or the vestibular nerve, the disorder is called a peripheral vestibular disturbance and may be differentiated from a central disturbance, which originates from malfunction of the vestibular nuclei or the higher central neural pathways.

PERIPHERAL VESTIBULAR DISTURBANCE

1. Diseases Accompanied by Cochlear Symptoms

a. Ménière's disease

In order to diagnose Ménière's disease, the following conditions must be satisfied. Vertiginous attacks accompained by nausea or vomiting occur repeatedly and the recurrence of the attack is associated with the exacerbation of cochlear symptoms. In most cases, unilateral sensory-neural hearing loss is documented by audiometry, and the alternate binaural loudness balance test (ABLB) or Békésy's audiometry shows the recruitment phenomenon. Peripheral vestibular disturbance is confirmed by equilibrium function tests. All cranial nerves except for the VIIIth nerve are unimpaired. X-ray of the temporal bone and otoscopic findings are normal. The Wassermann test reaction is negative (refer to Case Studies–1, p. 138).

b. Acute labyrinthosis (or vestibulopathy) with sudden deafness

The vertiginous attack is accompanied by unilateral severe deafness. The process of exacerbation is so rapid that it is also called an apoplectic form of Ménière's disease. This attack, however, may have a different pathogenesis from Ménière's disease and is thought to be caused by a virus infection or insufficient blood circulation in the inner ear (refer to Case Studies–4, p. 144).

c. Drug intoxication

Hair cells of the inner ear are sensitive to the toxicity of drugs such as salicylates, quinine, streptomycin, kanamycin, and some other antibiotics. Labyrinthine function is influenced by intoxication, and cochlear and vestibular symptoms may be produced.

d. Virus infection

Herpes zoster oticus (Hunt's syndrome), presumably caused by viral inflammation of the geniculate ganglion, is accompanied by facial paralysis and auditory and vestibular symptoms. Mumps deafness is rarely associated with vertigo.

e. Specific inflammation

Syphilitic labyrinthitis has Ménière-like symptoms. A positive Wassermann's test may differentiate between the two diseases. Cogan's syndrome, sarcoidosis and other so-called collagen diseases may exhibit evidence of vestibulo-cochlear involvement.

f. Acoustic trauma

The relation between intense noise and vertigo is known as Tullio's phenomenon. Sensory deafness caused by acoustic trauma is often accompanied by dizziness or unsteadiness. The audiogram may show a characteristic high tone notch (C_5 dip).

2. Vertigo Accompanied by Otitis Media (Vertigo Secondary to Ear Surgery)

Vertigo and disturbed equilibrium are often found in cases of otitis media, specifically in cholesteatoma otitis. In these cases, the following causes should be considered: perilabyrinthitis, labyrinthine fistula, serous or purulent labyrinthitis, and endocranial complications (cerebellar abscess) (refer to Case Studies–2, p. 140).

A peripheral vestibular disturbance is also common after ear surgery. The semicircular canal fenestration operation or stapedectomy for otosclerosis especially may be accompanied by long lasting vertigo.

3. Vertigo Induced by a Cervical Cause

When transient episodes of vertigo are induced by turning the head and neck, a cervicovestibular syndrome (HOZAWA, 1973) may be suspected. When the head is rotated and the neck extended, the vertebral artery can be compressed by a spondylotic spur. This mechanical pressure on the vertebral artery stimulates the perivascular sympathetic nerve plexus and induces vasoconstriction of the blood vessels of the inner ear, resulting in a peripheral vestibular disturbance. In such cases, spondylosis deformans or kinking of the vertebral artery can often be demonstrated by X-ray studies. The cochlea is not usually affected in this syndrome (refer to Case Studies–5, p. 146).

If vertebral arterial flow is compromised by chronic cervical osteoarthritis, transient vertigo accompanied by focal transient neurologic deficits due to medullary ischemia may be experienced.

4. Vertigo without Cochlear Symptoms

Vestibular neuronitis (DIX-HALLPIKE, 1952) and positional vertigo of the benign paroxysmal type belong to this category (refer to Case Studies–3, p. 142). The etiology of the first is obscure but the latter is thought to be caused by an otolithic lesion and is characterized

by transient nystagmus and vertigo which is induced by a specific head position. Acute viral labyrinthitis and epidemic vertigo, which is sometimes accompanied by brainstem encephalitis, are usually included in this category.

5. Vertigo of Unknown Etiology

There are a considerable number of cases, in which the causes of vertigo cannot be elucidated, although the results of equilibrium function tests and the absence of evidence of neighboring brainstem deficits are strongly suggestive of a peripheral vestibular disturbance. These include patients complaining of repeated attack of vertigo without accompanying auditory symptoms or a single attack of vertigo accompanied by auditory symptoms. We suggest that a tentative diagnosis of "aural vertigo of unknown etiology" be assigned to these patients.

CENTRAL VESTIBULAR DISTURBANCE

1. Cerebellopontine Lesion

a. Acoustic tumor

When unilaterally progressive deafness is accompanied by symptoms such as equilibrium disturbance, unilateral headache, and facial paresthesia, an acoustic tumor should be suspected. The main findings of the neuro-otological examination are as follows: 1) Retrocochlear deafness is verified by hearing tests: 2) hypoexcitability or loss of excitability in the affected labyrinth is observed by the caloric test and 3) gaze and positional nystagmus are characteristic. Bruns' nystagmus is often observed in the "brain tumor" stage. Positional nystagmus is mainly of the direction-changing type (NYLÉN's type I) and vertical or diagonal nystagmus can be elicited during the positioning test. If the disease has progressed, examination of the gait may reveal staggering, reeling or lurching. As a rule, the gait deviates towards the affected side. The spontaneous arm-tonus reaction may also be found as the arm on the affected side sinks down when the patient is asked to close his eyes and hold his arms out in front of him. A characteristic change in the optokinetic nystagmus pattern test can also be observed, but the so-called "reversion phenomenon" is not revealed by self-recording cupulometry. This phenomenon is observed exclusively when a labyrinthine lesion is present (refer to "The rotation test", p. 80).

Cerebellar and cranial nerve (V, VI, VII, IX) symptoms and signs are observed in the "brain tumor" stage. Morphological changes of the internal auditory canal or the pyramis are often demonstrated by X-ray (refer to X-ray Studies, Fig. 95, p. 121).

b. Other diseases

All tumors or pachymeningitis in the cerebellopontine angle display symptoms similar to acoustic tumors.

2. Lesion in the Brainstem ro the Cerebellum

a. Vertebral-basilar insufficiency

Symptoms and signs resulting from interference with the blood flow in the vertebral-basilar artery and its branches vary according to the site of the stenosis or occlusion, but do not always correspond exactly to any anatomical defect which may be demonstrable by neuroradiology or neuropathology. Rather, dysfunction results from the physiological defect in blood flow which is dependent upon the collateral circulation, blood pressure, heart rate and many other variables. For example, occlusion of one vertebral artery may or may not result in any noticeable central nervous system dysfunction.

The most common manifestations of vertebral-basilar insufficiency are transient dizzy spells, drop attacks, paresthesias of the face (especially perioral), or of one side of the face

and body, decrease in vision, double vision, slurred speech, and ataxia. If a history of recurrent episodes of vertigo is accompanied by such transient neurologic complaints, the physician should be alerted that dysfunction of the central nervous system is indicated. Vertebral-basilar insufficiency is, of course, most common in the population of patients over 50 years old (refer to Case Studies–8, p. 152).

b. Tumor

Pontine tumors, if localized to one side of the brainstem, cause contralateral hemiplegia and paralysis of the ipsilateral cranial nerves. These nerves are often interrupted in their intramedullary or intracranial course causing manifestations of peripheral nerve disease such as muscle weakness accompanied by atrophy. The Vth, VIth, VIIth or VIIIth nerves may be involved. If the tumors are intramedullary, dysconjugate eye movements and gaze nystagmus are common.

Cerebellar tumors give rise to different symptoms and signs depending upon the locus of the lesion. Gaze nystagmus and positional nystagmus may be observed with midline or paramidline tumors while hemisphere tumors usually produce defects limited to the limbs. Arteriovenous malformation may occur in the cerebellar midline or hemisphere and produces symptoms accordingly. If the tumor is calcified it may be noted on skull X-ray but the diagnosis is usually verified by angiography or pneumoencephalography.

c. Platybasia and other congenital abnormalities

In platybasia (basilar impression) pressure on the brainstem and cerebellar structures with compression of the basilar artery produces numerous symptoms and signs, among which gait disturbance and vertical nystagmus are characteristic. A congenital defect such as Arnold-Chiari malformation should also be considered in these cases.

d. Spino-cerebellar degeneration

Gaze nystagmus, positional nystagmus (direction-changing type), and positioning nystagmus (vertical type) are common. The characteristic findings of the optokinetic pattern test are demonstrated (refer to Case Studies–9, p. 154).

e. Multiple sclerosis

Multiple sclerosis may cause various ocular symptoms including gaze nystagmus, spontaneous or positional nystagmus (vertical or horizontal), and gaze palsy such as internuclear ophthalmoplegia long before the occurrence of other neurologic symptoms.

3. Cerebral Lesion

a. Tumor, abscess, meningitis

b. Epilepsy

Ablation of the temporal lobe due to tumor or abscess and inflammation in the subarachnoid space can produce vertigo or disturbance in equilibrium. Vertigo also occurs as an aura of epilepsy or as a seizure per se. In such cases it is necessary to examine the possibility of coexistence of peripheral and brainstem disorders.

VERTIGO AND DIZZINESS CAUSED BY HEAD INJURY

These cases can be divided into five groups (HOZAWA, 1961).

1. Group I

Peripheral vestibular disturbance is caused immediately by head injury. Labyrinthine fracture, labyrinthine hemorrhage or concussion belong to this group. In such cases the following symptoms are found: Hypoexcitability of the unilateral labyrinth is accompanied

by ipsilateral deafness. Ispilateral facial palsy can often be observed. Bleeding from the ear is common. Fracture of the labyrinth is confirmed by X-ray.

2. Group II

The occurrence of peripheral vestibular disturbance is delayed some weeks to months after head injury. Ménièriform disorder or positional vertigo of the benign paroxysmal type after head injury and the cervicovestibular syndrome due to traumatic spondylosis deformans belong to this group.

3. Group III

Neither central nor peripheral vestibular disturbance is confirmed by neuro-otological examination, however the patient complains of dizziness or tinnitus. In such cases, neurosis or malingering may be suspected.

4. Group IV

Central vestibular disturbance occurs without any evidence of accompanying cerebrospinal lesions. In group IV the abnormalities can only be demonstrated by the neuro-otological examination.

5. Group V

Central vestibular disturbance is accompanied by other cerebrospinal symptoms. Not only neuro-otological but also other neurological examinations show pathological findings. Utilizing X-ray tests (pneumoencephalography, angiography etc.) and electroencephalography, abnormalities can often be observed.

MISCELLANEOUS

Since vertigo and dizziness encountered in other conditions such as polycythemia rubra vera, sickle cell disease, diabetes mellitus, cardiovascular diseases, carotid sinus syndrome etc. could be both central and peripheral vestibular disturbances, it is difficult to classify them.

Recent Progress in Vestibular Research

Chapters on anatomy and physiology are not included in this book as our emphasis was placed on the description of procedures and interpretation of the neuro-otological examination. However, an attempt has been made to cover some recent advances concerning the basic aspects of peripheral and central vestibular mechanisms.

The parts of the inner ear related to the equilibrium function of the body consist of the three semicircular canals and the otolith organs, the utricle and the saccule. The receptor cells of the semicircular canals are stimulated by angular acceleration of the head, which causes movement of the cupula endolymph system and leads to deflexion of the cupula and of the sensory hairs. The action of linear acceleration or gravity causes dislocation of the otolithic membrane and the resultant bending of the sensory hairs stimulates the receptor cells in the otolith organ. However, the role played by the hair cell, the cupula or otolithic membrane, and the endolymph in the process of such mechano-electric stimulus transduction is not entirely clear despite a number of recent investigations.

The cupula of the semicircular canals has been classically viewed as a swinging door sitting in the ampulla. Recent evidence suggests that this may not be so. HILLMAN (1972) reported that the cupula actually has a circumferential attachment to the ampullar wall

and thus forms a diaphragm across the ampulla. He envisioned that the cupula moves like a sail with its center having maximum motion. Compression of the semicircular canal beyond physiologic limits (such as could occur in head trauma) might result in detachment of the cupula at its apex, a kind of safety valve mechanism. The cupula could then assume the classical swinging door appearance. OMAN and YOUNG (1972) in a theoretical consideration of the motion of the cupula find that the pressure gradients across the ampulla resulting from angular acceleration would result in extremely minute deviations of the cupula. Thus gross swinging door type motion would be unneccessary to explain the physiologic range of cupular motion.

Ultrastructural observation of the vestibular sensory epithelia of mammals and birds has demonstrated two types of sensory cells, i.e. the flask-shaped type I cells and the cylindrical type II cells (WERSÄLL, 1956). These cells have a marked difference in their contacts with afferent nerve endings; large chalices are formed around type I cells and smaller club endings are formed in the synaptic region of type II cells. In addition to afferent fibers the vestibular labyrinth is innervated by efferent fibers (PETROFF, 1955; WERSÄLL, 1956; ENGSTRÖM, 1958; GACEK, 1960). The connection of the efferent fibers to the receptor cells is also different for the type I and II cells. The efferent fibers can be demonstrated at the level of the brainstem (ROSS and CORTESINA, 1965). Thus the existence of a closed feedback loop between the labyrinthine receptors and the vestibular nuclei or reticular formation has been suggested.

The sensory hairs on the outer surface of the hair cells are divided into the kinocilium and the stereocilia. The kinocilium located in the periphery of the hair bundle is the longest and the stereocilia are arranged in order of decreasing height away from the kinocilium. Such an asymmetric arrangement of the hair bundle on each cell surface has suggested a characteristic organization in each sensory area of the labyrinth (LOWENSTEIN and WERSÄLL, 1959). For example, the kinocilium in the cristae of the lateral semicircular canal is regularly found on the utricular side, and in the cristae of the vertical canals on the opposite side. This may be related to the difference in directional sensitivity of the cupular movement between the lateral and the vertical (anterior and posterior) canals. However, functional significance of the detailed structures including that of the efferent system is still not clear.

Recently HILLMAN (1969, 1972) has proposed a mechanism to explain how motion of the labrinthine cilia can result in a modulation of vestibular nerve activity. The stereocilia which stand on a rigid cuticular plate cannot move vertically but are free to slide at their distal ends. The single kinocilium however stands over a notch in the cuticular plate but is attached to the stereocilia by filamentous processes at its distal end. Thus, when a force is directed toward the kinociliary end of the hair cell a plunging motion results in producing an inward deformation of the cell membrane at the kinociliary base. A force in the opposite direction would result in an outward rounding of the membrane. This deformation of the notch membrane could produce the conductance changes which are thought to modulate the transmitter release and modify VIIIth nerve activity.

Vestibular impulses from the labyrinthine receptors reach the vestibular nuclei and are transmitted to various portions of the nervous system such as the eye muscle motor nuclei, the spinal cord, the cerebellum, and the reticular formation. On the other hand, numerous afferent projections from other parts of the central nervous system converge onto the vestibular nuclei. Thus it is certain that the vestibular nuclei act not only as a single relay station but rather as an integrator of the activities from the periphery and from other sensory structures.

The anatomical and functional organization of the vestibular nuclei consists of four main