Development of Clinical Neurootological Network Diagnostics During the Last Three Decades

Claus F. Claussen
Forschungsinstitut der Gesellschaft zur Erforschung von Geruch-, Geschmack-, Gehör- und Gleichgewichtsstörungen e.V., Bad Kissingen, Germany

HISTORICAL DEVELOPMENT OF NEUROOTOLOGY

Since approximately 100 years ago, neurootology has been developed in medicine at the boundaries separating otorhinolaryngology, ophthalmology, neurology, and other medical subdisciplines. The science of neurootology addresses the diagnosis and treatment of the various diseases in the field of sensology (comprising the neurological aspect of otorhinolaryngology in diagnosis and treatment). It constitutes the systematic examination and treatment of diseases of the cranial senses. Therefore, it also includes the basics of neurosensory pathology and physiology.

With regard to the special complaints of patients affected with various ideological topical, traumatic, infectious, toxic, or systemic and underlying diseases, neurootology has centered around the symptoms of vertigo, dizziness, giddiness, nausea, hearing loss, tinnitus, and taste and smell disorders. Consequently, specific neurootological diagnostic approaches have been combined with a specific neurootological therapeutic approach.

In 1914, Viennese otolaryngologist Robert Bárány was awarded the Nobel prize for his pioneering examinations of nystagmus and the functions of the vestibular equilibrium. The ophthalmologist Ohm [1,2] in 1924 established the basis of optokinetic nystagmus analysis. Viennese neurologists Spiegel and Sommer, who published a seminal work on the topic in 1931, have named the entire subject area ophthalmo- and otoneurology. Earlier, in their very extensive multivolume manual of the neurology of the ear (1924–1929), otologists G. Alexander and H. Brunner and neurologist O. Marburg [1–5] stated:

... This manual's value lies in helping the doctor by assembling all publications about the complete field of sensory physiology together with everything that he must know for his work. The information is presented from a common viewpoint. We are called upon to accomplish an urgent task, of which the most basic part consists of compiling the arduous studies reflected in the otologic, neurological, and ophthalmological literature. Progress in these fields formed the strongest motive for us labyrinthologists to publish a manual of the neurology of the ear.

This book will help the clinician by explaining the application of anatomical and experimental research in this field. It is almost superfluous to note that the scientific examinations described have been possible only under full consideration of the neurological and otological aspects of the statoacoustic nerve. Any pathological anatomical examination of the hearing organ would be incomplete if it did not consider the total inner-ear organ up to the central area of the nuclei of the nervus statoacusticus.

In Vienna, we have formed an institute for the study of normal and pathological anatomy and physiology of the central nervous system. We also would very much welcome the establishment of an institute for normal and pathological anatomy and physiology of the sense organs. Such an institute would ensure more complete and rapid progress in the field of neurology of the ear...

In the former U.S. Army Air Corps (now the U.S. Air Force) and in the National Aeronautics and Space Administration, neurootology was promoted during and after World War II (1943–1976) by the very important work of Dr. Ashton Graybiel. Originally, Graybiel was a specialist in internal medicine. However, owing to the many scientific problems encountered in human air and space transportation, he narrowed his medical interests to a special application of neurootology in aviation and aerospace exploration. His work in
turn stimulated many other theoretical and clinical disciplines of medicine.

NEUROOTOLOGY: LABEL FOR A NEW MEDICAL SPECIALTY

As in other fields of modern medicine (e.g., the development of rheumatology from orthopedics and internal medicine), neurootology as we know it today developed from a combination of several older medical subspecialties. In its earliest incarnations, neurootology was established under three different labels: otoneurology, the subspecialty combining neurology and otology from the perspective of the latter discipline; neurootology, a specialty emphasizing neurology but originating from otology; and neuroophthalmology, which began as an ophthalmologic field viewed from neurological perspectives [6–20]. The prerequisites for developments in each of these arenas was medical systematics, the pathophysiological and pathological features of disorders of the cranial senses, including specific symptoms presented by affected patients. Such symptoms as vertigo, giddiness, dizziness, nausea, hearing loss, tinnitus, and taste and smell deficits are among those considered in these fields of study. Also included are such other related complaints as headache, forgetfulness, flight of thought, sleeping diseases, and pain. Any of these symptoms may occur individually or in combination (so-called multisensory complaints).

In its therapeutic aspect, neurootology—which was born of otorhinolaryngology—took two different routes. One route was toward neurootosurgery [10,13,19,21–24], whereas the other was a broad development toward conservative neurootological treatment [10,11,13,14,17,22,25–29]. The latter was possible only after World War I and was enhanced after World War II, when advances in medicine provided increasing knowledge about modern pharmacotherapy of the most varied human functional failures and disorders.

The title specialist in neurootology has been assigned for only approximately three or four decades, yet currently, worldwide, first chairs and special clinical complexes for the new medical specialty of neurootology have been established in Europe, America, and Asia.

THE LANGUAGE OF NEUROOTOLOGY

In neurootology, a language has developed that encompasses its own terms for describing functional approaches in diagnosis and therapy. The modern concept of noninvasive cranial sensory pathway analysis requires a modern, well-adapted, and concise lingual description of the failures found at various levels within the network. Our reports and diagnoses must be subjected to a new descriptive verbal standard. Because these neurootometrically induced and developed methods are embedded in the developments of our modern information age, it is fitting that the language applied to this science is borrowed from the language used to describe modern information technology [1–8,10–20,22,30–36].

By carefully evaluating the facts, our clinical neurootological team was able to conclude that the cause of some “diseases” could only be hardware failures of the underlying structures, whereas in other diseases, the cause was surely software-based. Hence, we have adapted and transferred into medicine these two terms—software and hardware—from modern information technology [7–20,22,23,26,29,31,33,37–39]. For instance, an acoustic neuroma, a schwannoma-type tumor of the eighth nerve, is a typical so-called morphological hardware disorder. Conversely, motion sickness in a normal person is a pure software disorder.

PRACTICAL NEUROOTOLOGICAL WORK

In its everyday application, modern neurootology deals primarily with the examination and treatment of disorders of the cranial senses. The main emphases lie in the fields of hearing and balance disorders and, less frequently, smell and taste disorders. Diagnostic breakthroughs were made in these fields by complete checking of the pathway systems, from the sensory receptors up to the cortical processors and sensory end projections. The objective and quantitative investigations, which are based on measurements of various types of reactions, are summarized under the label neurootometry. Tests within this specialty are arranged in groups. Equilibrimetry represents the measurement and recording of equilibrium functions [6–19,26,31–33,39–51]; audiometry, the measurement and recording of hearing functions [5,7,13–18,19,24,29,31,49,52–54]; gustometry, the measure of taste functions [15,18,19,36,55,56]; and olfactometry, the measure of functions of smell [9,15,18,19]. By measuring these functions, we gain new insights into quantitative, numerical, and charting ranges of normal and pathological states, which in turn lead to a system of disease definitions.

The results of modern neurootometric testing of sensory pathway functions are plotted on charts or recorded as numerical scores. This system provides objective descriptors of vestibular balance functions, which are checked biometrically from the inner-ear receptors through the brainstem up to the human cortex. The functional measurements are transferred as digital results into typical configurations of overall characteristics (e.g., butterfly, kite, and L-charts). Human pathological
Development of Neurootological Network Diagnostics

CONCEPT FOR CLINICAL NEUROOTOLOGICAL FUNCTIONAL NETWORK ANALYSIS

The concept of neurootological functional network analysis is based on a body of knowledge, which has been growing for more than 150 years, about the sense organs, their neurological connections through the brain nerves to the brain, and the pathways within the brain up to the cortical projections [1-5,7,9,11,13,16,17,19,30,34,36-38,52,57-61]. Long before the evolution of neurootology, much clinical research already had been completed (1906-1911) about the symptomatology of typical failures at the various levels within these functional pathways, failures that also have been demonstrated morphologically in anatomy. The well-known hierarchy of causal relationships between diagnosis and therapy must not be forfeited in this field of medicine. Therefore, throughout the recent decades, our neurootological research group at Würzburg and Bad Kissingen has made dedicated efforts to improve comprehensibility and reproducibility of causal diagnostics of the various diseases affecting the sensory network of the head [6-13,15-21,26,32,33,39-41,44-48,51,55,56].

Specific analytical methods have been developed to examine malfunctions of the balance-regulating system, assessing the various movements of expressive sensorimotor target outputs as indicators for either normal or pathologically altered functions. These methods ultimately led to the development of electronystagmography (ENG) in both its analog and digital forms [6-8,10-13,15-20,27,29,31,32,35,37,44-46,48,50,51,64]. Head and body movement failures are recorded by craniochoriography (CCG) [7,10,11,13,15,17-19,21,22,26,39,43,47,60-62]. Cortical brain functions are recorded by further developments of electroencephalography (EEG) through brain evoked potentials [9,14,18,19,33,40,49,52-54,57-59]; from this, a modern objective and quantitative audiometry could be devised. Additionally, we now use such imaging techniques as magnetic resonance imaging, computed tomography, and single-photon emission computed tomography, especially in tinnitus [24] or in diagnosing whiplash brain injury [19].

In the classic otolaryngologic fashion, patients with symptoms that are considered neurootometric are analyzed primarily by the recording of a medical history, interrogative exploration by psychophysical testing, and inspection of the sense organs in the ear, eye, nose, mouth, and larynx. Then typical objective and quantitative recording procedures are applied. Currently, neurootology supports medical intuition by charts, scales, and tables for quantitative test result comparisons with normal values.

Neurootometry and Equilibriometry

CCG was first designed and applied as a photooptical recording method for head and shoulder movements in 1968 by Claussen, mainly in a floor projection from above. To obtain such a recording, a patient’s head and shoulders usually are marked by light bulbs [7,11,13,15,17-19,26,39,43,47]. The recording of the light tracings is performed by means of an instant camera. Several exposures can be superimposed on the same film. For the quantitative evaluation of head and shoulder tracings, we then superimpose a relational polar coordinate system at the head’s height. The test chart, recorded by the camera, appears similar to a radar image. Additionally, movements of the head and shoulders can be compared during various tests of standing [60], stepping [61,62], walking, and other intracorporeal movements.

Currently, especially via digitized ultrasonographic computed CCG [63], head and shoulder movements can be recorded and evaluated permanently through a three-dimensional computer image (e.g., in normal position and in maximal flexion or maximal extension with related head rotations to either side) [19]. For diagnosing various balance diseases, we have combined the very rigid Romberg standing test [60] with the very sensitive Unterberger-Fukuda stepping test [62]. The Unterberger-Fukuda test displays the results of human
reorientation during locomotion, to which we have added the neck flexion-extension rotation (NEFERT) test for intracorporeal movement analysis [19,63].

All these tests provide investigators with typical reational patterns that then can be sorted as indicators of peripheral vestibular spinal or central brainstem or cortical pathological findings.

The modern ultrasonographic computed CCG chart delivers an instant test document that can be visualized and related directly to a test situation and an expected normal reaction. The document is easily understood, and the test involves very little effort on the part of patient or doctor. Likewise, medical aids and staff require little education to perform the test, and affected patients will cooperate readily. An entire measurement of the three regular CCG tests—standing, stepping, and head-neck motion—including quantitative evaluation and charting takes only 4 minutes.

**Electrocardiography**

For measuring vegetative reactions related to the so-called nausea complex, our neurotological teams at Berlin, Würzburg, and Bad Kissingen have used electrocardiographic (ECG) recordings simultaneously with ENG recordings for nearly 33 years [8,13–19,27]. The equilibriometric impact of ECGs is evaluated by changes in cardiac rhythms under modulation of vagal or sympathetic tonus. ECG recordings also are evaluated for cardiac dysrhythmias and extrasystoles.

**Objective Electroencephalographic Recordings and Related Techniques**

Owing to the technique of EEG, neurootology has a tool for inspecting functions within the cerebral pathways and in the end projections at the cortical surface. Thus, our measurements and observations apply throughout the functional processing within the brain pathways. The electrodes are positioned on the scalp, and the EEG reading is obtained according to classic EEG electrode mountings, amplifications, and recordings. However, modern computer technology has introduced the possibility of combining averaging methods with special topographic color display procedures. As a result, we have come to use vestibular evoked, acoustical evoked, visual evoked, and olfactory evoked brain potentials. Now, many test results can be displayed also in an interactive morphometric and functional chart system known as brain electrical activity mapping (BEAM) [9,14,17,19,29,33,40,49,52,57–59].

Sequentially dynamic EEG mapping is a new, high-resolution imaging procedure for investigating spontaneous brain activity and brain pathway activity under the influence of sensory stimulations from acoustic, vestibular, taste or smell, visual, and other sensors. Multichannel EEG permits derivation from the scalp of graphic online charts of EEG frequency behaviors or electrical voltage relations that can be displayed and printed by computers. These dynamic charts then offer the time course of topographically located patterns of cortical responses with respect to a defined stimulus. These most recently devised tools have provided a new approach to studying cortical sensory functions, particularly as they relate to tinnitus [9,19,29,49,52].

In our Würzburg University neurootological laboratory, we use vestibular acceleration stimuli on a rotatory chair for provoking vestibular evoked brain potentials. In patients with tinnitus or whiplash injury, we can demonstrate dramatic cortical reactional pattern changes under experimental stimulatory vestibular stress.

**Special Neurootometric Tests**

Now that neurootology has identified its new descriptive tools for functional analysis (e.g., ENG, CCG, ECG), and has successfully applied EEG of spontaneous and provoked patterns (evoked potentials, BEAM) to the study of neurootological dysfunction, we must define a functional analytical network that provides us with typical responses to a broader variety of neurootometrically designed tests. The most important and regularly used of these tests in our neurotometric toolbox are as follows:

- **Inspection:** Ear, eye, nose, mouth, pharynx, larynx, face, and neck, including sonography of the sinuses and computed rhinomanometry for patency of the nasal ducts [10,13,15,18,19].
- **Audiometric functional investigations:** Pure-tone audiometry; speech audiometry; reflex audiometry (including middle-ear impedance testing); measuring of acoustic dynamics, including the discomfort threshold measurement; tinnitus masking; transitory evoked otoacoustic emissions; computer-based objective hearing test using short-latency acoustic brainstem evoked potentials; and computer-based objective hearing test by late-evoked cortically evoked acoustic potentials of the cortical projection of the hearing pathways [14,15,18,19,29,31].
- **Equilibriometric functional investigations:** Recording and measuring of spontaneous nystagmus (open and closed eyes, dark or semidark room) with ENG; monaural caloric test of the vestibular ocular reflexes with bithermal stimulation, polygraphic ENG recording, and quantitative evaluation via the butterfly device; binaural pre- and
Development of Neurootological Network Diagnostics

postrotatory testing of the vestibular ocular system, with linear stimulation during the rotatory intensity damping test and evaluation through the rotatory intensity damping test L-scheme chart; vestibular stimulus response intensity comparison for finding vestibular recruitment or decrements, or other dynamic pathological processes; testing of the retinoocular system by ocular pendular tracking and by optokinetic nystagmus testing (free-field optokinetic and ENG recording); computer-based EEG analysis of visually evoked brain potentials; vestibular vegetative testing by recording of vestibular cardiac reactions through a simultaneous ECG; testing of the vestibular spinal system by head-body scheme analysis, with CCG recording to intracorporal movements and head and neck movements; standing test (CCG-Romberg); stepping-test CCG of the locomotor type (Unterberger-Fukuda); CCG of active head and neck rotation, with yaw, roll, and pitch analysis of the head with respect to the body and testing of the type of neck flexion, extension, and rotation; and vestibular evoked potentials with BEAM on the rotary chair (recorded vestibular evoked brain potentials) [6-8,10-21,23,26-28,31-33,35,39,41-48,50,51].

- **Doppler sonography of the major brain-supplying arteries**: Olfactometry; semiquantitativeolfactometry by target olfactography; olfactory evoked potentials with recording (BEAM) [15,18,19].
- **Gustometry**: Chemogustometry by five-component chemogustometry; electrogustometry by incremented-impulse electrogustometry [15,55,56].
- **Special neurootometric tests**: Complex equilibriometric tests (e.g., calorization pendulum interference test), cyclogram of habituation, and other tests used for special aspects of individual cases [7,11,13,16,17].

By means of these functional pathway analysis tests, we can verify many of the symptoms of our patients. In verifying them, we also can use the changes in the symptoms as a target for treatment by applying the functional failure descriptions elaborated by our test analyses. The tests allow the interrelation of positive or negative functional findings with typical morphometric (i.e., anatomic) relations to functional pathways within the circuits of visual, vestibular, hearing, proprioceptive, and other sensory systems [7,13,16]. We start from the sensors, pass through the brainstem and the cerebellum up toward the cortex of the telencephalon, and move back toward the motor drives of eye, head, neck, and body. Neurootological investigative methods are used routinely in conjunction with classic clinical methods, such as special inspections and other quantitative otorhinolaryngologic investigations (e.g., microscopy with television recording, rhinomanometry, radiography, magnetic resonance imaging, computed tomography).

### Special Neurootological History-Recording Procedures

Patients seek the help of doctors to cure special symptoms and are the best source of information about those symptoms. All patients first need the opportunity to voice their complaints, which then are recorded in a written history. Usually, two types of history-recording procedures are applied.

First a biographical history is requested, to which the doctor must listen carefully. This portion of the history is the patient’s description of symptoms in personal terms. Aside from this, we use a more extensive systematic neurootological case history (known as NODEC or NOASC), which is completed exclusively by examining doctors during the workup of a patient. The specific case history tool that we developed consists of a standardized case history questionnaire that leads an examining doctor from issue to issue. Such issues include specific details provided by a patient about headaches, and the like. Then we try to characterize any complaints related to nausea, such as sweating, palpitations, malaise, vomitus, and collapse. Additionally, we question for vertigo-releasing mechanisms (e.g., turning the head, bending, getting up, gazing to the side, gazing down or up). The duration of disease (e.g., the duration of a single vertigo attack) also must be ascertained. Through such questioning, we seek to uncover such additional symptoms as tinnitus, hearing loss, and visual disturbances (e.g., double vision, amaurosis, oscillopsia) and to assess the cranial senses (e.g., taste, smell, and trigeminal and facial nerves).

As regards background pathological processes, we ask questions that address trauma and neurological, otologic, metabolic, cardiovascular, and other disorders. Patients also are asked about current or former treatments. Among these treatments might be oncologic treatments and cardiovascular therapies, use of pain relievers, specific tinnitus or vertigo treatments, surgery, physiotherapy, and wearing of hearing aids.

Usually, the questionnaire ends with a patient’s self-rating of his or her follow-up. In this phase, each pa-
Table 1. Examples of Functionally and Morphologically Provable Diseases Related to Vertigo, Nausea, Tinnitus, or Hearing Impairment in a Topographically Ordered Scheme

<table>
<thead>
<tr>
<th>Functional Topodiagnostic Area</th>
<th>Diseases</th>
<th>Functional Topodiagnostic Area</th>
<th>Diseases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inner-ear receptors</td>
<td>Acute hearing loss</td>
<td>(Brainstem)</td>
<td>Platybasia</td>
</tr>
<tr>
<td></td>
<td>Acute vestibular loss</td>
<td></td>
<td>Prolonged ischemic neurological deficit</td>
</tr>
<tr>
<td></td>
<td>Capsular otosclerosis</td>
<td></td>
<td>Specific infectious diseases (tuberculosis, syphilis)</td>
</tr>
<tr>
<td></td>
<td>Complete inner-ear infarction</td>
<td></td>
<td>Status dysraphicus (Bonnevie-Ullrich syndrome, Klippel-Feil syndrome, Nielsen syndrome, Pierre-Marie’s disease)</td>
</tr>
<tr>
<td></td>
<td>Congenital syphilis</td>
<td></td>
<td>Stenosis of the aqueduct (aqueduct syndrome)</td>
</tr>
<tr>
<td></td>
<td>Perilymphatic fistula syndrome</td>
<td></td>
<td>Stenosis of the posteroinferior cerebellar artery</td>
</tr>
<tr>
<td></td>
<td>Hereditary inner-ear disorders (Pendred syndrome, Down syndrome, Mondini defect)</td>
<td></td>
<td>Syndrome of the bridge bonnet (Gasperini syndrome)</td>
</tr>
<tr>
<td>Inner-ear malformation</td>
<td>Intoxication with aminoglycosides</td>
<td></td>
<td>Syndrome of the edge of the clivus</td>
</tr>
<tr>
<td></td>
<td>Labyrinthine commotion</td>
<td></td>
<td>Slow-brainstem syndrome</td>
</tr>
<tr>
<td></td>
<td>Lermoyez syndrome</td>
<td></td>
<td>Arnold-Chiari syndrome</td>
</tr>
<tr>
<td></td>
<td>Ménière’s disease</td>
<td></td>
<td>Cerebellar abscess</td>
</tr>
<tr>
<td></td>
<td>Otitis interna of viral or bacterial etiology</td>
<td></td>
<td>Cerebellar atrophy</td>
</tr>
<tr>
<td></td>
<td>Otogenic tinnitus</td>
<td></td>
<td>Cerebellar heredoataxia (Nonne-Marie syndrome)</td>
</tr>
<tr>
<td></td>
<td>Retinitis pigmentosa (morbus Usher)</td>
<td></td>
<td>Cerebellar infarction</td>
</tr>
<tr>
<td>Eighth cranial nerve</td>
<td>Vascular inner-ear infarction</td>
<td></td>
<td>Cerebellar tumors</td>
</tr>
<tr>
<td></td>
<td>Temporal bone fracture</td>
<td></td>
<td>Alzheimer’s disease</td>
</tr>
<tr>
<td></td>
<td>Acoustic neuroma</td>
<td></td>
<td>Apoplexy</td>
</tr>
<tr>
<td></td>
<td>Meningitis</td>
<td></td>
<td>Behavioral disorders</td>
</tr>
<tr>
<td></td>
<td>Vestibular neuritis</td>
<td></td>
<td>Brain edema</td>
</tr>
<tr>
<td></td>
<td>Vestibular neuropathy</td>
<td></td>
<td>Chronic-toxic encephalopathy</td>
</tr>
<tr>
<td></td>
<td>Tumor metastases</td>
<td></td>
<td>Cortical atrophy (hunger or trauma)</td>
</tr>
<tr>
<td>Brainstem</td>
<td>Syndrome of the anterior inferior cerebellar artery</td>
<td></td>
<td>Cortical perfusion diseases</td>
</tr>
<tr>
<td></td>
<td>Anterior sinus cavernous syndrome</td>
<td></td>
<td>Cortical tinnitus</td>
</tr>
<tr>
<td></td>
<td>Apoplectic form bulbar paralysis</td>
<td></td>
<td>Cortical trauma</td>
</tr>
<tr>
<td></td>
<td>Arnold-Chiari syndrome</td>
<td></td>
<td>Creutzfeldt-Jakob disease</td>
</tr>
<tr>
<td></td>
<td>Basilar impression</td>
<td></td>
<td>Cysticercosis</td>
</tr>
<tr>
<td></td>
<td>Basilar insufficiency</td>
<td></td>
<td>Cysts</td>
</tr>
<tr>
<td></td>
<td>Basilar meningiomas</td>
<td></td>
<td>Dementia</td>
</tr>
<tr>
<td></td>
<td>Brainstem trauma</td>
<td></td>
<td>Depression</td>
</tr>
<tr>
<td></td>
<td>Brainstem intoxication (carbon monoxide, phenol, dioxin, etc.)</td>
<td></td>
<td>Dysplasia of the brain</td>
</tr>
<tr>
<td></td>
<td>Caudal syndrome of the red nucleus (Benedict paralysis, peduncular bonnet syndrome)</td>
<td></td>
<td>Encephalitis</td>
</tr>
<tr>
<td></td>
<td>Cervicoencephalic syndrome</td>
<td></td>
<td>Epilepsy</td>
</tr>
<tr>
<td></td>
<td>Dorsocaudal syndrome of the bridge bonnet (Foville syndrome)</td>
<td></td>
<td>Foreign bodies in the brain (bullets, clips, bone debris, etc.)</td>
</tr>
<tr>
<td></td>
<td>Dorsolateral oblongata syndrome</td>
<td></td>
<td>Foster-Kennedy syndrome</td>
</tr>
<tr>
<td></td>
<td>Encephalitis</td>
<td></td>
<td>Fractures of the skull</td>
</tr>
<tr>
<td></td>
<td>Garcin syndrome</td>
<td></td>
<td>Hereditary brain disorders</td>
</tr>
<tr>
<td></td>
<td>Gradenigo syndrome</td>
<td></td>
<td>Hydrocephalus</td>
</tr>
<tr>
<td></td>
<td>Hemiplegia alternans abducentis (Raymond syndrome)</td>
<td></td>
<td>Hysteria</td>
</tr>
<tr>
<td></td>
<td>Hemiplegia alternans inferior (Gubler-Miller paralysis)</td>
<td></td>
<td>Insula syndrome</td>
</tr>
<tr>
<td></td>
<td>Alternating oculomotor hemiplegia (hemiplegia superior, Weber’s paralysis)</td>
<td></td>
<td>Intracranial mass displacement</td>
</tr>
<tr>
<td></td>
<td>Interpeduncular syndrome (ventral syndrome of the foot of the brain peduncle)</td>
<td></td>
<td>Loss of cerebral efficacy</td>
</tr>
<tr>
<td></td>
<td>Lamina quadrigemina syndrome</td>
<td></td>
<td>Multiple sclerosis</td>
</tr>
<tr>
<td></td>
<td>Lateral oblongata syndrome (Wallenberg’s syndrome)</td>
<td></td>
<td>Parkinson’s disease</td>
</tr>
<tr>
<td></td>
<td>Multiple sclerosis</td>
<td></td>
<td>Postconcussional syndrome</td>
</tr>
<tr>
<td></td>
<td>Olivary dysgenesis</td>
<td></td>
<td>Psychoorganic syndrome</td>
</tr>
<tr>
<td></td>
<td>Olivopontocerebellar ataxia</td>
<td></td>
<td>Regional perfusion disturbances</td>
</tr>
<tr>
<td></td>
<td>Other brainstem tumors</td>
<td></td>
<td>Schizophrenia</td>
</tr>
<tr>
<td></td>
<td>Posteroimferior cerebellar artery syndrome</td>
<td></td>
<td>Temporal epilepsy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Temporal lobe tumors</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Thalamus syndrome</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Spasmodic torticolis</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Trauma to the temporal lobe with or without hematoma</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Whiplash brain injury</td>
</tr>
</tbody>
</table>
tient is asked to estimate whether his or her symptoms have remained unaltered for a long period or whether they have changed for better or worse.

NEUROOTOLOGICAL THERAPY

After the type of neurootological network lesions has been diagnosed and the descriptors for the functional failures have been established, the examining neurootologist must choose a therapy best suited for curing an affected patient or, at least, for relieving and diminishing the recorded complaints. Currently, we have five main approaches for treating neurootological disorders:

- **Pharmacotherapy**: For vertigo, dizziness, hearing loss, tinnitus, taste and smell changes, and other disorders
- **Physical therapy**: Through prostheses (hearing aids, tinnitus maskers, etc.)
- **Physiotherapy**: Such as vertigo- or tinnitus-training measures
- **Psychotherapy**: For phobias, anxiety-related vertigo, tinnitus, and the like
- **Neurootosurgery**: For tumors of the eighteenth nerve, posttraumatic states, and the like

ETIOPATHOLOGICAL CONSIDERATIONS IN NEUROOTOLOGY

In preceding publications, we have described neurootological approaches for the diagnosis of and therapy for various neurosensory disorders that can be described functionally [7,10,11,13,14,17,21–23,27–29]. On the basis of our experience with more than 100,000 patients over 33 years, we are able to differentiate systematically approximately 300 diseases related to neurootological symptoms. From this host of diseases, we have extracted a catalog of disorders that either are frequent or may serve as models of the underlying pathological findings of which we have a well-established knowledge (Table 1). However, we hasten to point out that this list was compiled mainly from the perspective of functional neurootological diagnoses, and many of these diseases also have a morphological or a psychometric component.

As stated in earlier sections of this article, functional lesions that have been described in the neurootologically defined network currently can be verified and localized by modern neurootometry. Clinically, however, we now must build a bridge between the functional neurootological identification of disorders within a topographically defined neuroanatomical network and a modern neurootometry-based and -monitored therapy. Hence, the structures of the sensory organs, the cranial senses, and the pathways and projections within the brain must be matched with functional indicators. Formerly, the pathological processes in these neuroanatomical structures of the sensory receptors, the pathways through the brain, and the cortical end projections of the senses were established by autopsies. Table 1 lists disorders (e.g., such related signs as vertigo, nausea, tinnitus, and hearing loss) that either comprise the complete spectrum of these subjective symptoms together with objective findings of lesions or exhibit only single components of what has been described. Included are typical examples of diseases defined in our medical books. For composite diseases with simultaneous damage in several sensory pathways, we now use the descriptor multisensory neurootological disorders.

EXPECTATIONS FOR NEUROOTOLOGY’S FUTURE

Neurootology’s development is parallel to and more or less synchronous with trends found in modern medicine. A special challenge for both modern medicine and neurootology lies in the fact that our knowledge of the diagnoses of and specific therapies for many diseases is expanding and may change rapidly in increasingly shorter periods. In contrast to many newly established medical branches that repeatedly abolish boundaries established around a medical subspecialty when the mother discipline has grown too rapidly and, therefore, has become incomprehensible as a single subject, neurootology increasingly exhibits its capacity to be holistic, synergistic, and synthesizing, embracing all knowledge about the neurosensory diseases, regardless of the various medical specialties in which these diseases often are addressed. Thus, as Alexander, Marburg, and Brunner [1] expressed as early as the time of Bárány, neurootology meets an urgent demand for uniting whatever knowledge is necessary for better understanding the pathophysiology of the cranial senses. As such, neurootology can be viewed as a crystallization point within the permanently changing world of medical facts and figures.

Both diagnostic and therapeutic standards are adapted constantly to meet the steadily increasing demands of patients suffering from vertigo, nausea, hearing impairment, tinnitus, and other disorders. Most new neurootology patients today are the elderly in developed countries, where life span expectations are climbing rapidly. In these groups of older persons, we observe linear or even progressively increasing numbers of presbyvertigo, presbyataxia, presbytinnitus, and presbycusis cases. Another major group of patients seeking help from neurootologists is composed of persons who were exposed to trauma (e.g., whiplash injury or intangi-
cation with modern chemicals or solvents) leading to chronic-toxic encephalopathy.

A new and unique opportunity lies in the systematic combination of neurootological differential diagnosis with constantly improving therapeutic approaches. Most patients present to the neurootologist with multisensory neurootological disorders. Neurootology, therefore, forms the basis for a modern international concept of clinically applied sensology. The field of neurootology is strengthened by classic literature in the field and by rapid information exchange on the Internet, in such regular journals as the Neurootology Newsletter and the International Tinnitus Journal, and (in the field of neurootology) in seminars and publications under the auspices of the Neurootological and Equilibriometric Society and other neurootological organizations such as Neurootologisches Forschungsinstitut der 4-G-Forschung e.V.

REFERENCES


