Paparella: Volume II: Otology and Neuro-Otology

Section 1: Diagnosis of Disorders of the Ear

Chapter 1: Otologic Diagnosis

Michael M. Paparella, Michael S. Morris

In this opening chapter of Volume 2, time-proven principles of diagnosis in the field of otology are discussed, as well as recent research and how it affects concepts of training, treatment, and rehabilitation. The reader is referred to chapters in which newer developments in the profession are discussed (unless a specific volume number precedes the chapter number, this volume is meant). Finally, concepts of the interrelation of otopathologies, the interactions of diseases of the middle ear and inner ear, and the interplay between diagnostic and therapeutic measures are discussed.

Recent Developments in Otologic Research

With basic clinical and technological research and development, otology has become and is becoming more sophisticated. Basic research and the basic sciences, as encompassed in Volume I, are essential to understanding normal structures and functions, and then to understanding the etiologies and pathogeneses leading to pathologic conditions and the pathophysiologic symptoms of the many diseases pertinent to otology. The following chapters of this volume document the many advances to which clinical research has contributed. Clinical research results from application of basic research to the patient and the patient's problem (disease), as well as to clinical experience and astute observation of problems and their solutions in patients, sometimes through serendipitous applications or by trial-and-error methods.

Technological research and development has led to better tools for diagnosis, treatment, and rehabilitation in otology. Examples of diagnostic aids seen in Volumes I, II, and IV include computerized tomography (CT), magnetic resonance imaging (MRI), immunobiological studies, and audiologic and vestibular studies. Examples of aids for treatment include better operating microscopes, and lasers. Other innovations and developments contributing to better otologic treatments include shorter hospital stays for otologic procedures, and more procedures being done in ambulatory surgery and surgicenter environments.

The field of neuro-otology, including surgery for acoustic tumors and trans-temporal bone section of the vestibular nerve, is covered in this volume. The methods of skull-based surgery have broadened the clinician's approach to previously unresectable neoplasms affecting the temporal bone and related regions. Newer techniques have been developed in tympanoplasty and mastoidectomy (eg, intact-bridge tympanomastoidectomy, the IBM procedure). Prostheses for structures of the middle ear and reconstructive techniques for the middle ear also assist modern otologists in helping their patients. We are on the threshold of new developments that will allow us to treat diseases of the inner ear such as deafness and vertigo, something that has not hitherto

been possible.

Examples of developments in rehabilitation include smaller, less visible, more effective hearing aids and assistive listening devices. Other new developments in otology, such as cochlear implants and bone-conductive implants and, possible in the future, implantable hearing aids, apply to the areas of both treatment and rehabilitation.

Although must recent progress is derived from clinical contributions and research in otology and closely related fields, otology is indebted to many other fields for contributions. Examples include contributions from the basic sciences and from related disciplines.

Education and Training

Considering the above, it is no wonder that it is difficult for the otologist or otolaryngologist to maintain current knowledge and skills. Training and education in such an expanding field is a lifelong career goal. While residencies may need to be modified to accommodate expansion of specializations within the field, current American Board of Otolaryngology (ABO) requirements provide for various options for concentrated study in otology. Fortunately, there are many postgraduate otologic courses and symposia available to the otologist who wishes to refresh his knowledge and skills and to remain current.

During the past several decades, the otolaryngologist has been evolving and expanding from the somewhat limited "ear, nose, and throat doctor" to what we prefer to term the "ear, head, and neck physician", as capsulized in these volumes, even though otology still constitutes a substantial volume of work within the profession. Although otolaryngology is practiced and defined along a continuum, the critical "definition" still occurs in the universities and other medical centers in their undergraduate and graduate programs. Training programs have been evaluated and upgraded to serve educational needs with staffs of appropriate size and an appropriate patient-care mix (Lierle, 1968a). The ABO has kept pace with the continuing evaluation and development of otolaryngology by recommending (in 1978) that residency be of five years' duration (Work, 1982); it was subsequently adopted to include three or four years of clinical training (Work, 1979).

While the ABO continues to discuss the issue of subspecialty certification, in recognition of the expansion of the field and the limited number of cases available to help every otolaryngologist learn to do everything, certain program directors are recommending flexibility during the years of residency training. After exposure to general otolaryngology, a "weighted" or "tracking" experience can be provided to the resident during final stages of training (Bailey, 1975). Thus, in a given residency program, one could elect a track to become a head and neck surgeon, an otologist, or a generalist in ear, nose, and throat (Schuknecht, 1979). For example, sparse resources for clinical study of the ear could be reserved for training those choosing to become "otologists" in their fifth year (Schuknecht, 1981).

Otology as a graduate discipline is appropriately taught in all United States

otolaryngologic residency/training programs. Of the more than 100 programs in the United States, however, no two are alike in opportunities for clinical otologic training. Some programs are located in larger cities, where there may be a surplus of otolaryngologists in practice, while others are found in smaller cities with a more rural environment. Some programs are strong in head and neck surgery but weak in otology; a recent tabulation of directors of training programs in the United States revealed that the majority of directors have expertise in head and neck surgery, rather than in otology. Some traditional programs do have strength in otology, defined by having experienced faculty otologists and sufficient clinical material for training, but in the United States contemporary otology is also practiced, and important contributions are made by groups at private otologic centers.

Because of the patterns of concentration of patients and teaching otologists, it is commonplace for aspiring otologists to seek supplemental one-year fellowships in a program strong in otology or neuro-otology, for a concentrated period of training and to gain additional experience in this field. These fellowships exist in traditional teaching centers in universities and also in the private sector. Opportunities for clinical otologic training depend on the volume and kinds of clinical cases available to the training program and on the ability of the faculty to treat and attract patients to medical centers and to use cooperatively private resources for patient care in community facilities.

During the period of change within the field, several factors will affect the practice and study of otology, as they affect the economics of medicine in general and otology in particular. The relatively increased concentrations of otolaryngologists in urban rather than in rural areas, and in relation to the overall population in the United States is a factor; there are 8000 otolaryngologists in the United States, and in the Japan, with a population half the size of that in the United States, there are also 8000. Another factor is that indigent patients have all but disappeared from certain training programs, and with a variety of mechanisms for payment available to patients in the United States, and even the likelihood looming of a national law regarding health insurance, all patients will become private patients supported by second- and third-party mechanisms for payment and using new and growing delivery systems such as health maintenance organizations (HMOs), independent practice associations (IPAs), and preferred provider organizations (PPOs).

The opportunity for clinical study alone, however, even when patients are available in abundance, does not in itself provide a suitable atmosphere for otologic training. There have been many instances in which residents have learned poor otology in institutions where full-time or part-time faculty were not available to provide guidance in thought and deed. The trainee/learner needs to have structured guidance in the use of head and hands in learning to manage patients with otologic disease.

Historical Perspective

We are first reminded of the philosophy of training during the Lempert School of the 1930s and 1940s. A select cadre of otologists was trained by Dr. Lempert, who performed

surgical fenestrations for otosclerosis; however, the vast number of remaining otolaryngologists were not otherwise trained to do this more sophisticated kind of otology.

In the 1950s, Wullstein and Zöllner introduced concepts of tympanoplasty. In the same decade, contributions by Rosen and Shea led to the development of stapedectomy. These two major contributions in the field of otology were assisted by the introduction and use of the operating microscope during the 1950s. Other noteworthy otologists such as Schuknecht contributed to a basic understanding of otology as well as clinical otology, whereas in the field of neuro-otology, William House helped to pioneer. During this important period, there were many other contributions to the development of otology by notable otologists such as Cawthorne, Shambaugh, House, Boies Sr, and others.

The development of procedures for tympanoplasty and stapedectomy stimulated a proliferation of interest in these new methods of managing ear problems, and this interest quickly permeated our residency/training programs. The net result of this is that many more otolaryngologists are now being trained to perform microscopic procedures on the ear. During the 1960s and on into the 1980s, there continued to be widespread dissemination and refinement of techniques and knowledge in the field of otology.

In a sense, then, the pendulum has swung from one extreme, when the majority of otolaryngologists were prohibited from performing current procedures, to the present state, when today's resident expects to learn and do everything, including everything in the field of otology. As in most cases, the pendulum needs to rest somewhere in the middle, so that proper care of patients can result from properly trained physicians with experience in managing otologic problems.

Concepts in Training

Progressive clinical training of an otologist starts with the first year of residency and continues until complete retirement from caring for patients. Important attitudes and habits of self-development are often (but not always) initiated and influenced during the formative years of residency. Residencies and/or fellowships provide the environment, opportunity, and role-models; however, only the individual physician can provide the motivation and sustained effort for lifelong learning. The years of residency provide an opportunity for graduate apprenticeship; as always, active involvement (doing) is superior to passive involvement (watching). By performing as well as observing methods of diagnosis and treatment, the individual physician develops a sense of confidence that provides a good foundation for further learning (Paparella, 1975).

If all the art, knowledge, and skills involved in practicing modern otolaryngology were placed in a box, that box would be minuscule in comparison with the box required to accommodate the unmet needs for diagnosis and treatment of diseases of the ear, head, and neck. Residency programs in otolaryngology should strive, therefore, to teach the ability to be at least a consumer of (or better yet, a contributor to) new clinical knowledge reported in the literature.

Clinical research and clinical training are inseparable. At the least, one is obligated to understand research if one is to read current otolaryngologic journals, and it is our conviction that training in research enhances the clinical capabilities of the otolaryngologist.

Medical Versus Surgical Otology

The role of the physician-teacher is to teach others to be physicians first, not just surgical technicians. A good head is more important than "good hands". The otologist as a thinking physician is interested in the biologic history of disease, including its etiology and pathogenesis, and is aware, through a continuing assessment of the literature and through other means of keeping abreast, of the best means of management. This is true for interest in all otologic diseases, recognizing that relatively few patients seen initially in a physician's office later come to surgery. The otologist should be an internist as well as a surgeon; should be interested in the diagnosis of all otologic problems, in sensorineural as well as conductive hearing loss, in vertigo, and in infectious problems; and whenever possible should treat the patient conservatively. Even for patients requiring surgical procedures, the thought processes of the physician should be actively involved in initial diagnosis, preoperative considerations, the surgical procedure itself, the postoperative healing period, and the period of rehabilitation.

Benchmarks (Including Dissection of Temporal Bones)

It should be kept in mind that there is great variation among training programs. Some provide three years of otolaryngologic training, others four. Some have excellence in otologic training; others have weakness in this area. Establishing uniform benchmarks, therefore, that will apply to all otologic programs is not necessarily appropriate. General objectives for a program, or specific benchmarks that list desirable objectives for each year of clinical training, will vary from program to program and will depend on the expertise of the clinical faculty and on the volume and nature of clinical resources for patient care.

Starting with courses in basic science and an introduction to basic principles of diagnosis and treatment, with an emphasis on pathogenesis, the individual otologist should have an increasing opportunity actively to evolve the ability to diagnose and treat patients during training, and also during continuing conferences and coursework. In general, a otologic resident should dissect at least 12 temporal bones, especially early in training, in order to develop manual dexterity and familiarity with the operating microscope. Ideally, this too should be a continuing process throughout the residency or fellowship and also later in life. It is important to practice using a technique thoroughly on temporal bones before an attempt is made on the living patient. An introduction to temporal bone dissection is discussed in Chapter 13 of this volume. After this experience has been acquired using temporal bones, then experience in surgical otology is appropriate.

We have found the two-operating room setup to be most useful in teaching otologic surgery: the staff surgeon works in one room, allowing the resident or fellow to initiate the case in the second room; then, surgeon and resident switch back and forth, providing an opportunity

for the resident to participate and to obtain on-site supervision. Feedback and the ability to monitor a resident's or fellow's performance are important to an otologic residency or fellowship (Harker, 1978).

Qualities emphasized in the maturing otolaryngologist include (1) clinical judgment, (2) ability to communicate with patients, (3) ability to communicate with colleagues, (4) moral and ethical values, (5) acceptance of responsibility, and (6) continuing responsibility.

The following implementing principles provide for a gradual, balanced progression in all the major areas of medical and surgical maturation during residency or fellowship: (1) factual information, (2) interpretative skills, (3) information-gathering ability, (4) problem-solving ability, (5) clinical judgment, and (6) surgical skills.

Lifelong Graduate Student

After graduation from residency, the physician becomes a lifelong graduate student, at least until retirement from active practice. The word "practice" is intriguing when applied to medical practice; if practice is supposed to make perfect, then the physician who practices the art and science of otology hopes to achieve an unachievable state of perfection. This concept of private practice is in line with the concept of continuing or post-graduate education; more than anything, the continuing quest for knowledge and skills assures the highest possible quality of care to the patient.

There are abundant opportunities for continuing training. The American Academy of Otolaryngology - Head and Neck Surgery provides prime examples. Through the Academy, special educational opportunities are available. Excellent programs and courses are provided by universities, private groups, and various societies, and at many meetings held in the United States, and in many other countries. The field of otology does not seem to lack for selection in postgraduate opportunities for training.

Although it is important to teach principles or fundamentals of otology in training programs, and to allow residents or fellows opportunity to learn by doing, it is also important to instill in the student the concept of self-assessment or self-determination that should lead to continuing self-improvement. The physician should know his or her abilities and limitations and should be aware of the quality of care that can be provided in this area of practice. In many instances, difficult and special problems should be referred to a more experienced otologist for consultation or management, or both. Conscience should be the guide; various mechanisms for peer-review may also play a role. It is not an admission of defeat but rather sound responsibility to refer patients with problematic cases to others with particular experience with such cases. Good results for the patient, with complications minimized, should be the first concern.

In sum, the goal is to teach and to practice quality care. "Quality care" has many definitions. A useful one phrased by Walter McClure, director of a Minneapolis medical care "think tank", is (1) patient satisfaction, (2) outcome (result), and (3) innovation. The "art" of

medicine continues to supersede in magnitude, by far, the "science" of medicine. It is important not only to treat the disease but also to treat and support the patient. Managing the patient requires high ethical and moral standards and principles, and the time and ability to communicate with the patient in order to obtain "informed consent" regarding the patient's problems and the possible methods of management.

Diagnostic Concepts

Diagnosis

An otologist is a specialist who deals with diseases of the ear. As such, his or her role is the same as that of any physician dealing with any other organ-system in the body. The first function of the otologist is to diagnose the disease, to consider options of conservative medical management, and then, if indicated, to consider options of surgical management with the patient. After considerations of diagnosis and management follows rehabilitation, including use of hearing aids.

Too often, otolaryngologists will not attempt to diagnose many disorders of the ear. For example, many patients present with the various diseases causing sensorineural hearing loss. It is important in every instance to make a working diagnosis or to diagnose the disease causing the symptom of hearing loss or the finding of hearing loss as documented by an audiogram. The role of the otologist is to use the same approach for all the symptoms or problems of patients with otologic disease, including vertigo, tinnitus, pain, infection, pressure, and so forth.

We will elaborate specifically on the role of the otolaryngologist in managing hearing loss in the following paragraphs. The same principles apply, however, for all the other symptoms and diseases in the field of otology. Simply stated, the most important part of the diagnostic workup is usually the history, followed by physical examination, and then appropriate laboratory studies. The simplest laboratory studies should be considered to control costs and to make a diagnosis, preserving the more elaborate studies such as MRI for specific indications in patients.

The Role of the Otologist

The following description of the role of the otologist is based on loss of hearing as the patient's chief complaint or primary presenting symptom. Hearing loss is a symptom or a finding (if documented audiologically) of some otologic disease. The hearing loss may manifest as conductive, mixed, or sensorineural. Primary symptoms of diseases of the inner ear are hearing loss, tinnitus, and vertigo. Hearing loss and tinnitus arise from the front part of the inner ear (auditory labyrinth), which is related to hearing, while vertigo arises from the posterior portion of the inner ear (vestibular labyrinth), which is related to maintenance of balance and equilibrium.

The vestibular labyrinth (pars superior) develops embryologically and phylogenetically earlier than does the auditory labyrinth (pars inferior). It is axiomatic that older structures and organs appear to be more resistant to disease, whether developmental or acquired, than are newer

structures. Perhaps this is why more diseases affect the auditory labyrinth, causing hearing loss and tinnitus, than affect the vestibular labyrinth, causing only vertigo. Of course, there are many diseases causing both hearing loss and vertigo. It is also generally agreed that approximately 10 per cent of the American population (ie, 22 million persons) suffer from a significant problem with speech and/or hearing. It is obvious that a health-care problem of this magnitude has profound social and educational implications.

Every patient with a hearing loss requires first and foremost a medical diagnosis and, if possible, medical management. The only trained professional capable of performing this function is the otolaryngologist. There is a need for the otolaryngologist to expand his or her knowledge of hearing and to assume appropriate responsibility for the benefit of the patient. The other key professional is the medical audiologist. Hearing loss can be stable or dynamic, over time, and ideally every hearing loss requires the attention of both the otologist and the audiologist.

The respective roles of otology and audiology are well described in the fourth edition of *Hearing and Deafness*:

Otology is the medical and surgical specialty that deals with the organs of hearing and balance from the point of view of their diseases and the prevention or treatment of these diseases, as well as to safeguard the life of the patient. Otology is part of the field of otorhinolaryngology, that medical specialty which deals with diseases of the ear and head and neck. Its point of view is primarily biological. Audiology is a specialty that is concerned with the function of hearing, with strong emphasis on its educational and social aspects and of providing assistance, where appropriate, in the form of hearing aids.

Nevertheless the two specialties interact strongly and provide much mutual support. The otologist can do much to relieve conductive hearing impairment due to abnormalities or pathology of the external or middle ear, and can give relief medically to some disorders of the middle and inner ear. Audiology in its turn can provide the otologist with very useful information from several audiological tests, particularly those which are designed to locate the anatomical site of a lesion. Sometimes a differential diagnosis, such as that between an auditory neurinoma and Ménière's syndrome, is vital and may be greatly assisted by tests performed by the medical audiologist.

A final principle is that both fields share responsibility for the management of the individual patient, but decisions regarding otological management should always precede decisions on audiological management. Biological safety must be insured and function restored as fully as possible. On the other hand, surgical or medical intervention to improve or restore auditory function must be evaluated in terms of the auditory potentialities that remain as well as many local and general clinical considerations.

Hearing loss is a medical problem. Before a patient receives medical treatment or audiologic habilitation or rehabilitation, every attempt should be made to establish a definitive medical diagnosis or working diagnosis; that is, before we can properly manage a hard-of-hearing

patient, we should know what we are dealing with.

Too often the symptoms of hearing loss are described, categorized, and treated, while the problem per se or the underlying process leading to the symptoms is not treated. For example, if the patient is said to have severe sensorineural hearing loss with greater loss in the high frequencies, and there is markedly reduced discrimination considered to be of cochlear origin, this is useful information, but it does not diagnose the underlying disease that resulted in these findings. In *Dorland's Medical Dictionary* (1988), a disease is defined as "any deviation from or interruption of the normal structure or function of any part, organ, or system (or combination thereof) of the body that is manifested by a characteristic set of symptoms and signs and whose etiology, pathology, and prognosis may be known or unknown". Hearing loss can occur as a result of a problem in the peripheral or central auditory system (local) or as a local manifestation of some systemic disease such as hypothyroidism.

Federal legislative studies of this problem have been going on for years. An FDA regulation that became effective August 15, 1977 states that every patient in the United States in whom hearing loss is suspected must have medical clearance by a physician, preferably one specializing in diseases of the ear, prior to purchase of a hearing aid. Here we are discussing what is best for the patient from a medical point of view, not political or legislative. Nevertheless, this new law provides a challenge and opportunity for appropriate medical care and management for patients with hearing loss.

Early identification of hearing loss is the responsibility of the patient, the family, the school teacher, health professionals - in short, all of us. When a hearing loss is suspected, the patient should be seen by an otologist working with an audiologist. Every patient with suspected hearing loss or deafness requires a careful medical history and physical examination of the head and neck, but especially of the ear. Ancillary studies are selected as needed, but audiology is an essential aid in the diagnostic process. The vast majority of problems with hearing loss can be diagnosed and managed within the outpatient setting. The cost-effectiveness of an outpatient workup considerably surpasses that of studies done for the hospitalized.

The primary emphasis of the diagnostic workup is to rule out all possible extrinsic causes of deafness or hearing loss and, by the process of exclusion, to be left with an intrinsic cause, or deafness that is likely to be genetic. Extrinsic causes include infection, drugs, tumors, trauma, neurologic problems, congenital causes, metabolic causes, and so on, and there are literally hundreds of possibilities. Through otologic nd audiologic assessment, one identifies conductive, sensorineural, or mixed hearing losses. Conductive losses, in the main, can be treated and corrected through medical or surgical means. Diagnostic micro-otoscopy and therapeutic microsurgery are of immense benefit in this regard. As we learn more about sensorineural hearing loss, we are discovering which of these conditions can be managed medically.

In Chapter 40 and 41, details of the history-taking process for diagnosing sensorineural hearing loss are described. Usually, expensive testing (such as CT, electronystagmography (ENG), and so forth) is not necessary. The most important part of the diagnostic process, by far, is the

history. Second in order of importance is the examination of the ear and the head and neck, together with the audiometric configuration and other audiologic information. This preliminary workup leads to a definitive diagnosis or a working diagnosis for the majority of all sensorineural hearing problems. As outlined in Table 1 and as described in Chapter 40 in detail, the first consideration is whether the hearing loss is congenital or delayed; the next consideration is whether it is genetic or nongenetic.

Table 1. Classification of Deafness

Congenital Deafness	**Delayed Deafness**
Genetic	**Genetic**
1. Deafness occurring alone	1. Deafness occurring alone
2. Deafness occurring with other abnormalities (syndromes)	2. Deafness occurring with other abnormalities (syndromes)
3. Chromosomal abnormalities	
	Nongenetic
Nongenetic	
	1. Inflammatory diseases
1. Deafness occurring alone	2. Ototoxic poisoning
2. Deafness occurring with other abnormalities	3. Neoplastic disorders
	4. Traumatic injury
	5. Metabolic disorders
	6. Vascular insufficiency
	7. Diseases of the central nervous system
	8. Presbycusis.

Rarely, a patient with sensorineural hearing loss or deafness will require more elaborate studies that may be best accomplished on an in-patient basis. Detailed protocol for such a diagnostic workup was originally described by Meyerhoff (1977), and Meyerhoff and co-workers (1984) (see Table 2). Some of the indications for such studies include the following conditions:

1. The deafness is idiopathic and develops rather suddenly without even a working diagnosis.

2. There are systemic problems suggesting a regional disease - for example, deafness associated with metabolic diseases, diseases of the central nervous system, thyroid diseases, syphilis, kidney diseases, and so forth.

3. There is fluctuant hearing loss suggesting, for example, endolymphatic hydrops or perilymphatic hypertension or fistula.

4. Preliminary findings suggest a retrocochlear or central nervous system problem.

5. The patient develops sudden hearing loss, especially if the patient is seen soon after occurrence of the problem.

6. Additive or fluctuant hearing loss occurs in a patient with formerly stable sensorineural hearing loss.

It is helpful for the otolaryngologist to identify the hearing loss as stable, progressive, or fluctuant. It is possible for a progressive sensorineural hearing loss to become fluctuant years later, or a fluctuant hearing loss can become stable or progressive subsequently. Also, additive causes may ensue after the initial diagnosis. For this reason, it is important that all patients with a sensorineural hearing loss be seen periodically, preferably once a year or at least every two years, for follow-up.

Table 2. Diagnostic Protocol for Deafness

History (otolaryngologic and complete general)
Physical Examination (otolaryngologic - head and neck - and complete general, including cranial nerve assessment). Include consultations for
- Funduscopic examination
- Neurologic evaluation, especially of the cranial nerves
- Medical evaluation for vascular, collagen, or other systemic disease

Audiologic Evaluation (frequent re-evaluation will assess progression, improvement, or stability)
- Tuning fork tests: Rinne and Weber (others optional); use Barany's noise box when necessary (these tests are to be done by a physician to corroborate audiologic findings)
- Whispered voice and shout test (using Barany masking)
- Pure tone air- and bone-conduction tests
- Speech reception threshold tests
- Phonetic balance (discrimination)
- Modified tone decay test
- Loudness recruitment test - short-increment sensitivity index (SISI), alternate binaural loudness balance (ABLB) test, or both
- Impedance measurements to include acoustic reflex
- Auditory brain-stem response (ABR) audiometry

Vestibular Evaluation
- Romberg sign - tandem, standing
- Gait
- Spontaneous nystagmus (note direction and type)
- Positional test
- Caloric test
- Electronystagmography, using air-caloric testing method

Radiologic Evaluation
- Skull radiogram
- Chest radiogram
- Computed tomography (CT) scan

- Magnetic resonance imaging (MRI)

Cardiac Examination

- Vital signs (blood pressure, pulse, respiration, temperature)
- Electrocardiogram (ECG)

Hematologic Examination

- White blood count and differential count
- Hemoglobin
- Sedimentation rate
- Platelet count

Coagulation Studies

- Prothrombin consumption
- Prothrombin time
- Partial thromboplastin time
- Platelet count
- Definitive studies if either hypercoagulation or hypocoagulation is suspected

Renal Evaluation

- Urinalysis
- BUN
- Creatinine

Endocrine Evaluation

- Thiocyanate flush (for Pendred syndrome)
- Fasting blood-sugar determination
- ACTH plasma cortisol stimulation
- Protein-bound iodine determination
- Cholesterol and triglyceride determination
- Glucose tolerance test

Biochemical Studies

- Total protein
- Albumin
- Globulin

Serum Electrophoresis (optional, unless indicated by history or examination)

- Sodium, potassium, calcium, chlorine, carbon dioxide

Liver Function Tests (optional, unless indicated by history or examination)

Serologic and Immunologic Evaluation

- VDRL
- FTA-ABS
- Lupus erythematosus cell preparation
- Heteroagglutinin titer
- Antigen-specific cellular imune test for middle ear antigens
- Cryoprecipitation (quantitative and qualitative, C-1q binding assay)
- Total hemolytic complement assay (CH50)
- C-reactive protein

Lumbar Puncture

- Opening pressure

- Color
- Cell count and differential count
- Electrolytes
- Serologic studies
- Glucose
- Viral culture
- Culture for bacteria, fungi, and acid-fast bacilli

Viral Studies

- Acute specimens (should be obtained as early as possible within, but not after, 21 days of onset of deafness) of whole clotted blood (for culture and titer)
- Stool
- Washings from throat or nasopharynx, or both
- Cerebrospinal fluid
- Fluid from middle or inner ear, or both; under special conditions - such as tympanotomy for evaluation of a perforated round window - these may be available for viral culture and should be inoculated into prepared tissue-culture dist in the operating room
- Convalescent whole clotted blood (should be obtained for culture and titer between third and fifth week after onset of deafness and at least 14 days after obtaining acute specimen of whole clotted blood)

Exploratory Tympanotomy (indicated when spontaneous rupture of round window or other middle ear disease is suspected, or if perilymph is to be sampled for diagnostic purposes).

Multiple Otopathologies

It is axiomatic of medicine in general and of otology in particular that diagnosis of disease depends strongly on an understanding of the etiology or causative mechanisms that initiate the disease and of the pathogenesis, mechanisms, or processes that transpire between the cause and the end-state of the pathologic or pathophysiologic condition, usually manifested by symptoms, that brings the patient to the otolaryngologist's office. Pathologic findings, in general for medicine and particularly in the temporal bone for otology, remain perhaps the most significant correlate for diagnosis and therefore for treatment of disease (Paparella, 1978a). In a previous study of 1383 temporal bones from 713 patients, multiple pathologic lesions were systematically assessed. In 11 per cent, or 152 temporal bones, there was more than one pathologic finding. Males (60.5 per cent) had multiple diseases more commonly than did females (37.7 per cent). The most frequently occurring findings were otitis media (71.1 per cent), otosclerosis (43.4 per cent), endolymphatic hydrops (38.8 per cent), labyrinthitis (25.0 per cent), and cancer (24.3 per cent).

Pathologic findings, the end-result of etiology and pathogenesis, are important correlates in otologic diagnosis. Multiple coexisting pathologic conditions can have coincidental or causative relationships. The otolaryngologist should consider multiple pathology when diagnosing and treating diseases of the ear. As an example, there is evidence to suggest that otosclerosis may in some causative way predispose to Ménière's disease and that otitis media can cause sensorineural hearing loss.

It is difficult enough for the otologist to diagnose one disease in the ear; it behooves him, however, to consider other coexisting diseases, either occurring coincidentally or with a causal relationship. Also, there may be systemic diseases which are either causal or coincidental in terms of otologic problems. For example, syphilis is likely to have a causal relationship to labyrinthine symptoms, whereas cardiovascular disease, hypertension, and diabetes are likely to be coincidence in relationship.

Interaction Between Middle Ear and Inner Ear

There are many diseases in which there is interaction between the middle and inner ear. An increasing number of otologists and researchers are studying and documenting these relationships (Paparella et al, 1987b). As our understanding of these interactions improves, so will our ability to diagnose and treat otologic diseases. In the future, exciting opportunities will exist for sampling fluid from the middle and inner ear for diagnosis and for perfusion of the inner ear as therapy in selected cases. In this brief, summarizing overview, we attempt to categorize these clinical pathologic entities and make observations relative to diagnosis and treatment, with comments on the role of exploratory tympanotomy.

Table 3 documents otologic diseases in two categories: (1) diseases of the middle ear cleft with inner ear manifestations and (2) diseases of the inner ear with middle ear manifestations.

Table 3. Otologic Diseases

Diseases of Middle Ear with Inner Ear Manifestations	Diseases of Inner Ear with Middle Ear Manifestions
1. Congenital anomalies	1. Congenital anomalies
2. Trauma (including changes from pressure)	2. Trauma (including changes from pressure)
3. Infection / inflammation	3. Infection / inflammation
4. Tumors	4. Tumors
5. Granulomas	5. Otosclerosis
6. Ototoxic drugs	6. a. Ménière's disease (decompensated)
7. Cochlear implants	6. b. Ménière's disease with perilymphatic fistula
	7. Perilymphatic hypertension.

Diseases of the Middle Ear with Manifestations in the Inner Ear

Congenital Anomalies

Congenital anomalies in the middle ear cleft usually occur separately from anomalies of the inner ear because of the separate primordial and embryologic origins. Sometimes, however, developmental anomalies of both the middle ear and inner ear can occur together. Exploratory tympanotomy will often provide diagnostic information useful in evaluating these anomalies.

Obstruction of both windows can occur from congenital fixation of the stapes, aplasia of the oval or round window or of both, or a high jugular bulb that obstructs the round window (Paparella, 1980). If obstruction occurs in both windows, a sensorineural hearing loss will appear on the audiogram as a result of elimination of phase differential. A recent prospective study indicates that an anteromedially displaced lateral sinus, with associated reduction in Trautmann's triangle, is commonly seen in patients with Ménière's disease. This congenital developmental abnormality may, later in life, lead to endolymphatic malabsorption and symptoms and findings of Ménière's disease (Paparella and Sajjadi, 1989).

Trauma

Trauma through the middle ear can result in damage to the membranous labyrinth, leading to possible cochlear or vestibular damage and dysfunction. Iatrogenic trauma can result from stapedectomy surgery for otosclerosis (Paparella, 1966). Trauma may also result from penetrating foreign bodies, which can fracture and subluxate the stapedial footplate into the vestibule (Arragg and Paparella, 1964). Fractures of the temporal bone that include the promontory may serve as a source of entry for inflammation of the middle ear to pass into the inner ear. In addition, acoustic trauma to the ossicular chain can occur from a drill during atticotomy or tympanomastoidectomy (Paparella, 1962).

Trauma to the round window can inadvertently result from surgical procedures while removing a cholesteatoma or granulation tissue from the round window niche during tympanoplasty, or from damage with the myringotomy knife - for example, if the myringotomy is placed in the posterior inferior quadrant of the tympanic membrane. Aggressive irrigation during tympanomastoidectomy can damage an exposed, delicate round window membrane. Other forms of iatrogenic surgically induced trauma include all surgical procedures in the middle ear cleft that have the potential for causing labyrinthine damage. We have no better example, perhaps, in which to observe this than in the variety of methods of surgical treatment for intractable Ménière's disease. Labyrinthine damage can result from endolymphatic sac surgery, puncture of the dilated saccule (Cody's tack, Fick operations), cochleosacculotomy, crysurgery, and ultrasound treatment for Ménière's disease. Singular neurectomy for intractable benign postural vertigo can also be complicated by cochlear damage.

Changes in Pressure

Changes in barometric pressure such as that encountered in scuba diving or descent during air flight can result in a locking of the eustachian tube and a sudden negative change of atmospheric pressure in the middle ear cleft, leading to rupture of the round window and a perilymphatic fistula (Fee, 1968; Goodhill et al, 1973). Perilymphatic fistula can be associated with sensorineural hearing loss and may appear as part of the sudden deafness syndrome (Chap. 43), or perilymphatic fistula can be associated with vestibular symptoms, including imbalance, dysequilibrium, and vertigo.

Infection/Inflammation

For some time, we in our laboratory have been interested in sensorineural hearing loss occurring secondarily to otitis media. We find this to be a fairly common phenomenon and have conducted a number of laboratory and clinical studies to better understand this phenomenon. Otitis media, both acute and chronic, often associated with cholesteatoma and granulation tissue, can contribute to sensorineural hearing loss. Vestibular symptoms can also result from invasion of toxic materials either through a fistula of the horizontal semicircular canal (cholesteatoma) or through the oval or round window. It is well established that in the tympanogenic form of labyrinthitis, otitis media and its products can spread through the round window membrane to cause suppurative labyrinthitis (Turner and Fraser, 1928; Druss, 1929; Sprowl, 1931; Dean and Wolff, 1934; Paparella and Sugiura, 1967). The actual round window membrane, lying deep within the round window niche, can even be confused with a false membrane of the round window niche that is a mucosal superficial membrane (Nomure et al, 1983). While performing tympanoplasty, exploratory tympanotomy, or tympanomastoidectomy for otitis media, the round window is always carefully searched for.

If sensorineural hearing loss clinically appears to exist as a concomitant of otitis media, our policy is to pack Gelfoam into the round window niche in order to provide thickening of the membrane so as to protect against further episodes of contamination from otitis media. Recent temporal bone studies in patients with chronic otitis media indicate a thickening of the round window membrane in chronic compared with acute forms of otitis media (Sahni et al, 1987). Tracer studies show evidence that in chronic otitis media the thicker membrane is less permeable than the thinner membrane in acute otitis media (Schachern et al, 1987a). Gelfoam, which adheres to and thickens the membrane, was found to be the best grafting material if the round window membrane was intact (Schachern et al, 1987b). The thickened membrane served as a barrier to a tracer substance (Schachern et al, 1987b).

Clinicians and investigators have observed and described sensorineural hearing loss in otitis media prior to our interest in the subject (Turner and Fraser, 1928; Druss, 1929; Sprowl, 1931; Dean and Wolff, 1934; Hulka, 1941; Aberg, 1953; Gardenghi, 1955; Fricklinger, 1957; Verhoeven, 1961; Bluvshtein, 1963; Rauch, 1965; Thorburn, 1965). Many of our studies relating to this subject are referenced (Sahni et al, 1987; Schachern et al, 1987a, b; Yokoyama et al, 1968; Paparella et al, 1970a; Kawabata and Paparella, 1977; Paparella, 1977; etc).

An assessment of the round window membrane's role in the pathogenesis of disease requires an understanding of its normal detailed cytoarchitecture. Ultrastructural characteristics of the round window membrane have been studied in animals such as monkeys, guinea pigs, chinchillas, and cats (Goycoolea, 1987; Kawabata and Paparella, 1971) and in humans (Schachern et al, 1984). Authors have reported a relationship of cochlear loss to otitis media with effusion (secretory otitis media) in children (Arnold et al, 1977; Hutchings, 1978; Moore and Best, 1980; Munker, 1981; Aviel et al, 1982; Paparella et al, 1984a, b; Hellstrom et al, 1985; Dommerby and Tos, 1986). Studies and observations of this common phenomenon have documented sensorineural hearing loss in patients with chronic otitis media (Paparella et al, 1970a, 1984a, b), acute suppurative otitis media (Paparella et al, 1984; Morizono et al, 1985), and silent or subclinical otitis media (Paparella et al, 1980a, b, 1984a, b, 1986; Giebink et al, 1985).

The pathologic correlate appears to be serous labyrinthitis localized to perilymph of the basal turn, resulting in temporary threshold shift (TTS)(Paparella et al, 1972; Morizono et al, 1985). Over time, with continued exposure to otitis media, permanent threshold shift (PTS) or permanent sensorineural hearing loss can result, with subsequent apical involvement of lower frequencies. Studies in animals include tracer studies to assess the permeability of the membrane in otitis media and electrophysiologic evidence of cochlear losses in the basal turn in experimentally induced acute otitis media. In addition, animal and human studies have indicated that endolymphatic hydrops can be associated with or may result from otitis media, particularly chronic otitis media.

Occasionally, sonoinversion can result from a healed perforation of the tympanic membrane from previous otitis media. In such an instance, the round window membrane is exposed to and directly receives conducted sound, allowing the mobile stapes to serve the usual role of the round window membrane in phase differential. In this case, the traveling wave presumably goes in the opposite direction, and we have seen examples in which conductive sensorineural hearing loss is small or nonexistent. This natural observation has been created surgically by Garcia Ibanez in certain cases of tympanoplasty.

Anatomic-pathologic changes of the middle ear can be seen in certain children or adults with persistent otitis media. For example, patients may have atelectasis with the malleus markedly retracted and attached to, sometimes imbedded in, the promontory. In other patients, the promontory is very high, thus creating an obstruction to the protympanum and to the ventilation or drainage function of the eustachian tube. These anatomic-pathologic precursors can contribute to clinically manifest chronic otitis media including silent (subclinical) otitis media and can contribute secondarily to inner ear damage (Paparella, 1986). These anatomic problems can be compensated for by cutting the tensor tympani tendon, lateralizing the malleus, and grafting a deficient membrana propria of the tympanic membrane plus inserting a ventilation tube, to gain resolution of middle ear disease (Paparella and Jung, 1981).

Tumors

A variety of tumors in the middle ear, occurring either primarily or secondarily, can destroy the bony labyrinth and thus enter the membranous labyrinth. Clinically, tumors also have been seen to invade the round window niche and enter the labyrinth. We have seen several glomus jugular tumors which have invaded the round window niche, resulting in sensorineural hearing loss. The most common tumors that can cause labyrinthine damage, thus resulting in both cochlear deafness and vestibular symptoms, include (1) glomus tumors, (2) squamous carcinoma, and (3) adenocarcinoma (Adamas et al, 1971).

Granulomas

A variety of granulomas can involve the middle ear cleft, and as these granulomas expand they can cause damage to the labyrinth. Granulomatous diseases that have the potential for causing inner ear problems secondarily include the following: (1) tuberculosis, (2) cholesterol

granuloma, (3) eosinophilic granuloma, (4) nocardiosis, (5) Wegener's granulomatosis, (6) histoplasmosis, (7) polyarteritis nodosa, (8) sarcoidosis, and (9) syphilis.

Ototoxic Drugs

It has recently been observed in human and animal studies that ototoxic drugs (eg, Corticosporin) can cause cochlear damage by entering the round window membrane (Morizono and Sikora, 1982; Meyerhoff et al, 1983; Wright and Meyerhoff, 1984). The higher frequencies are the first to be affected by toxic agents present in the middle ear. High-frequency audiometry is essential, to assess not only problems of ototoxic drops but also sensorineural deafness resulting from otitis media and other forms of sensorineural deafness in which the high frequencies are involved. Ototoxic agents that easily penetrate the round window membrane have been used therapeutically for selective vestibulotoxic effect to treat vertigo; for example, streptomycin and gentamicin have been so used. Many studies have been described in which various agents instilled in the middle ear are recoverable in the inner ear. At all times, cochleotoxic effects are of concern.

Cochlear Implantation

We have now reached an era in which cochlear implantation has become a clinically acceptable method of rehabilitation or treatment of profound deafness in postlingually deaf patients. It is now fairly well established that cochlear implantation provides an enhanced opportunity for patients to receive sound-intensity and, to a lesser extent, speech discrimination. There are also studies describing the traumatic effect of inserting electrodes into the inner ear. At all times there is a need to balance the therapeutic effect of implantation with the traumatic effects of the insertion of such instrumentation into and through the scala tympani.

Diseases of the Inner Ear That Influence the Middle Ear

Congenital Anomalies

An enlarged cochlear aqueduct or patent modiolus can be a precursor of perilymphatic hypertension, labyrinthine membranous damage, and ultimately perilymphatic fistulas. Mondini's deformity and deafness can also be associated with aplasia of the oval or round window or drainage of perilymphatiic or even spinal fluid through both windows (Paparella, 1980).

Trauma (Including That From Changes in Pressure)

Horizontal fractures of the temporal bone will not heal by primary intention because of the unique characteristics of intrachondrial bone. Such nonunion of fractured sites can predispose to infection and cerebrospinal fluid otorrhea. Straining and heavy lifting (for example, from sports and exercise) can lead to increased spinal fluid pressure, increased perilymphatic pressure, and possible resultant perilymphatiic fistulae. The existence of precursory patency of the cochlear aqueduct and modiolus is a prerequisite.

Infection/Inflammation

Infection from bacteria or inflammation from viruses and other agents can cause changes not only in the endolymph but also in the perilymph that may result in endolymphatic hydrops, thus interfering with inner ear conductivity and phase differential. Perilymphatic inflammation may result in perilymphatic hypertension. Suppurative labyrinthitis has three stages: (1) acute, characterized by proliferation of polymorphonuclear leukocytes; (2) fibrous, characterized by fibroblastic proliferation, particularly in the perilymphatic spaces; and (3) labyrinthitis ossificans, characterized by the formation of bone throughout the membranous labyrinth (Paparella and Sugiura, 1967). If labyrinthitis ossificans is present, or possibly other changes resulting from membranous inflammation or infection, there will be lack of a round window reflex and interference of the traveling wave through the inner ear. This can be observed in some patients during exploratory tympanotomy. While there are rare indications (eg, labyrinthectomy), if one were to look with a microscope into the vestibule through the oval window, one could also see evidence of infection of the inner ear.

Tumors

Tumors of the middle ear are more likely to invade the inner ear. Tumors of the cerebellopontine angle are less likely to enter the labyrinth, and thus the middle ear. Metastatic tumors to the petrous apex and labyrinth can present in the middle ear. Meningiomas can also present as tumors in the middle ear cleft. Tumors that occupy the inner ear, once again, will result in loss of a round window reflex (Adams et al, 1971).

Otosclerosis

Otosclerosis is perhaps the best example of a labyrinthine disease with middle ear manifestations. Otosclerosis occurs within the bony labyrinth, its most important predilective site being the fistula ante fenestram. By virtue of its growing in the bony labyrinth, secondarily the footplate of the stapes is invaded and fixed, thus leading to classic conductive deafness. It is also well established, however, that otosclerosis causes sensorineural hearing loss, concomitant usually with conductive deafness. Moreover, otosclerosis can grow internally into the scala tympani so as to obstruct round window niche, once again interfering with phase differential and the round window reflex. We have observed a few patients who have had conductive losses even after successful stapedectomy. Upon revision, we could see no round window reflex, and therefore assume that in such patients the round window is obstructed on the scala tympani side, owing to otosclerotic bone formation in the scala tympani.

Immune Response in the Inner Ear in Experimental Stapedectomy

Through serendipity, distinct pathologic findings in the immune response of the inner ear, characterized by lymphoid follicles, were seen to develop after experimental stapedectomy in several animals (Paparella et al, 1967). A prospective effort was then made to reproduce these findings in animals that had received prior sensitization with homologous skin grafts and had then

undergone experimental stapedectomy. One-third of the animals successfully demonstrated the same distinct pathologic findings in the immune response of the inner ear (Gromer et al, 1974).

Ménière's Disease - Decompensated

Advanced Ménière's disease may result in endolymphatic hydrops that completely obstructs the vestibule owing to saccular hydrops as well as completely obliterating the scala vestibuli. We have observed clinically in such patients that not only may there be interference with phase differential but also the round window reflex may be absent in such patients.

Ménière's Disease and Perilymphatic Fistulas

A number of recent publications describe the coexisting occurrence of perilymphatic fistulas and Ménière's disease. There are a number of theories to describe such an occurrence, and indeed patients are now being treated clinically for both entities (Swift, 1985; Arenberg et al, 1986; Ganz, 1986; Parell and Becker, 1986; Seltzer and McCabe, 1986; Wieder, 1986; Allen, 1987; Nomura et al, 1987; Paparella, 1987).

Perilymphatic hypertension, a recently described clinical syndrome, is defined as a precipitous drop in hearing or as deafness, usually in a patient with congenital or acquired sensorineural hearing loss (Paparella et al, 1987a, b). It is often accompanied by physical exertion or trauma to the head. We have observed a small subset of patients who develop sudden deafness and bulging of the round window membrane, but without perilymphatic fistulas or perilymphatic drainage. After careful, innocuous paracentesis, followed by grafting, we have seen certain patients regain their hearing so as to be functional once again. Perilymphatic hypertension results in a bulging round window membrane. It is considered a precursor to perilymphatic fistula. Paracentesis of the round window membrane followed by grafting of collagen can result in restoration of hearing (Paparella et al, 1987b).

As with perilymphatic fistulae, a patent or semipatent cochlear aqueduct and modiolus are considered prerequisites for this condition. This observation helps support the concept that a perilymphatic fistula per se may not be the cause but can represent the end-result of events involved in certain forms of sudden deafness. An analogy might be acute otitis media resulting in a spontaneous perforation of the tympanic membrane or for which a paracentesis (or myringotomy) can be used to treat the disease. Furthermore, in addition to the inflammatory products that can enter the cochlea, secondary endolymphatic hydrops may ensue as a result of loss of perilymph (Nomura et al, 1987) or air can enter the labyrinth, resulting in dysfunction (Yanagihara and Nishioka, 1987).

The Role of Exploratory Tympanotomy

Patients who have diseases resulting in interaction between the middle ear and inner ear require consideration of, first, an appropriate medical diagnosis, then possible treatment, and finally rehabilitation including the use of a hearing aid (Paparella et al, 1987b). The most

important diagnostic workup includes the history, followed by otoscopic and physical examination and appropriate laboratory studies, including audiologic and vestibular tests and when necessary polytomography, CT scans, and MRI. Many findings in the middle ear, as well as findings of interaction between the middle ear and inner ear, can be determined (and sometimes treated) by exploratory tympanotomy, but not, for example, by CT scan. These include observed changes in details of soft tissues in the middle ear, ossicular abnormality, and mobility including fixation of the footplate; changes in the round window reflex and characteristics of the round window membrane; perilymphatic hypertension; and perilymphatic fistulas.

Examples of diseases of the middle ear with labyrinthine complications are more common than those from the inner ear having middle ear manifestations. From a pragmatic as well as a philosophic point of view, every time the otologist performs surgery for any disease in the middle ear cleft, in a sense he or she is concomitantly performing exploratory tympanotomy. That is, an otologist cannot be certain of what anatomic or pathologic changes exist until and unless they are observed directly under the microscope. Very often, patients who are scheduled for stapedectomy or tympanoplasty are found have some other disease process or unexpected pathologic condition. For example, certain patients who have intact tympanic membranes and persistent conductive or mixed hearing losses and other clinical evidence of otitis media are found to have chronic silent otitis media.

The classification or definition of otitis media is still a matter of controversy. We have modified the following simple classification from one recommended by a recent report by an international panel (Lim et al, 1985):

1. Suppurative otitis media

    a. acute
    b. chronic

2. Nonsuppurative otitis media (secretory otitis media or otitis media with effusion

    a. serous
    b. mucoid.

In this committee's report, chronic otitis media is defined as a condition associated with a perforation of the tympanic membrane with a history of otorrhea. This definition of chronic otitis media can be seen in most textbook references. However, an earlier study clearly demonstrated intractable pathologic findings in tissues in 81 per cent of cases studied in human temporal bones (92 cases) in the absence of perforation of the tympanic membrane or otorrhea (Meyerhoff et al, 1978).

Silent (masked) otitis media is a term that we originally employed to refer to pathologic conditions behind an intact tympanic membrane, which may be "undetected" or "undetectable", especially in chronic cases (Paparella et al, 1980b, 1984a). Silent otitis media has also been

described in experimentally induced otitis media in animals. Evidence has also been presented indicating that insidious labyrinthine changes such as sensorineural hearing loss (Paparella, 1981) or endolymphatic hydrops (Paparella et al, 1979) can be associated with or result from silent otitis media. Earlier, pathologic and clinical-management correlates for four clinical problems associated with silent otitis media were discussed as follows: (1) silent otitis media associated with meningitis in infants, (2) silent otitis media - the continuum, (3) silent otitis media - sequelae, and (4) chronic silent otitis media (Paparella et al, 1986).

Exploratory tympanotomy is a simple, cost-effective, direct, safe, useful means for diagnosing pathologic conditions in the middle ear and many of the interactions between the middle ear and inner ear. Indications for exploratory tympanotomy, as reviewed by a consensus of authorities, are listed in Table 4.

Table 4. Indications for Exploratory Tympanotomy

Exploratory tympanotomy can be used to diagnose and sometimes to treat (1) pathologic conditions in the middle ear and (2) interactive pathologic conditions in the middle ear and inner ear.

1. Pathologic Conditions in Middle Ear:

a. Conductive hearing losses suspicious for otosclerosis or other ossicular pathologic findings.

b. Objective tinnitus with suspected etiologic bases in middle ear.

c. Masses in middle ear requiring identification and/or biopsy.

d. Chronic otitis media unresponsive to medical therapy.

2. Interactive Pathologic Conditions in Middle Ear and Inner Ear: Diagnosis and, if possible, treatment of interactive diseases including infection, trauma, and perilymphatic fistula (PLF) or perilymphatic hypertension (PH). More specific indications:

a. Undiagnosed cochlear symptoms, fluctuating or progressive or sudden, including sensorineural hearing loss unresponsive to medical therapy, in which PLF or PH is suspected.

b. Undiagnosed vestibular symptoms unresponsive to medical therapy, in which PLF or PH is suspected.

c. (Rarely) patients presenting with strongly suspected PLF who require emergency exploratory tympanotomy (eg, patients post-stapedectomy or post-trauma).

3. Indications for Grafting of Round Window or Oval Window:

a. Observed perilymphatic fistula.

b. Suspected previous occurrence of PLF.

c. Deficient membrane in oval window or round window.

Exploratory tympanotomy is simple, cost-effective, direct, safe, useful means for diagnosing pathologic conditions in the middle ear and many of the interactions between the middle ear and inner ear. Indications for exploratory tympanotomy, as reviewed by a consensus of authorities, are listed in Table 4. We utilize three sizes of exploratory tympanotomy: small, medium, and large. A transcanal exploratory tympanotomy using a stapes-type flap can be utilized to explore the middle ear and its contents while assessing all of the variants including

changes in the round window. If, however, attic disease is anticipated, a small endaural exploratory tympanotomy will provide for access to this area with subsequent coverage with the initial flap; or, if one anticipates the possibility of mastoidotomy and/or mastoidectomy, either an intact wall or an intact-bridge tympanomastoidectomy (IBM), a large endaural exploratory tympanotomy can be done with exposure of the mastoid cortex. The nature of the clinical situation will dictate the type of exploratory tympanotomy.

As in all otologic surgery, the approach to a patient's specific problem should remain flexible. Experience illustrates potential differences between preoperative "assumption" and operative "facts". Surgical technique should always be flexible enough to accommodate the pathologic conditions identified intraoperatively. Using the stepwise methodology mentioned above will allow for identification and eradication of diseases as it is uncovered. The actual procedure or set of procedures selected will be directed by surgical findings rather than preoperative testing. The more flexible the approach, the better the treatment possible for pathologic conditions encountered.

Treatment of Otologic Disease

Treatment of otologic disease is based on the medical diagnosis. At all times, the goal is to institute conservative treatment. Medical treatment always precedes considerations of surgical treatment, if medical treatment applies. An important component of medical treatment is explaining the problem to the patient and supporting the patient psychologically. It is important to remember that the disease is to be treated, but also the patient requires management. Medical treatment for otologic disease is documented throughout the various chapters of this volume on otology.

Surgical therapy has indications for purposes of treatment and, as described earlier in the discussion of exploratory tympanotomy, occasionally for purposes of diagnosis. Once again, conservative surgical therapy should precede more radical surgical therapy to achieve desirable end results. If possible, procedures that may alter pathogenesis should be considered before destructive operations. This particularly applies to surgical treatment for Ménière's disease, as is discussed in Chapters 49 and 50. It should be stressed to the patient and in teaching that there are three equally important parts of surgical therapy: before, during, and after. Thus, even the best procedure may not have a good long-term result if the patient does not return for follow-up for maintenance of a good result. This particularly applies to surgery for chronic mastoiditis and chronic otitis media (see Chap. 32). The diagnostic thought process and judgment continues during surgery and during follow-up visits.

There are many examples of prevention of disease using conservative surgical therapy. The use of ventilation tubes can stop the continuum of changes due to otitis media, thus preventing further and more advanced disease (see Chap. 27). Methods of delivering otologic surgery are changing according to newly evolving health-care plans and because of changes in medical economics that affect medicine in general and otology in particular. For example, otologic procedures such as stapedectomy and tympanoplasty, which would have routinely

required inpatient hospitalization, are now performed safely by many physicians in a surgicenter or ambulatory care surgical facility. We have evolved from hospitalizing almost all of our otologic surgical patients to finding that many of these procedures can safely be done in an ambulatory setting.

While we are witnessing an evolution from inpatient to outpatient otologic surgery, other methods of delivering otologic care are changing also. It is traditional, for example, for an otologist to perform a procedure on one ear at a time. We know, however, that many children will have ventilation tubes inserted bilaterally, for example at the time of tonsillectomy and adenoidectomy. If the otologist is experienced, it may be in the patient's best interest to consider bilateral procedures for certain select cases (eg, myringoplasty) so as to reduce future hospitalization, risks of anesthesia, and so forth. As mentioned earlier, follow-up is important. If the patient lives a long distance from the hospital, new methods of delivering care will help to provide better care and better follow-up for such patients.

Rehabilitation for Otologic Patients

Rehabilitation for patients with hearing loss is generally a role that the otologist shares with the audiologist and, in some instances, the hearing-aid dispenser. Hearing aids are an example of rehabilitation in the field of otology. So are assistive listening devices. These are described in Chapter 9, 10, and 11. New procedures are described in this volume, including cochlear implants and implantable hearing aids. These two procedures seem to border on the areas of both treatment and rehabilitation. In the future we will have to consider modifying definitions of what constitutes treatment and what constitutes rehabilitation for the otologic patient.