

DISORDERS OF THE AUDITORY SYSTEM

Second Edition

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Foreword

Writing a book on disorders of the auditory system is a daunting task. Few individuals have the depth and breadth of knowledge to accomplish it effectively. But the authors of this volume, Dr. Frank E. Musiek, Dr. Jennifer B. Shinn, Dr. Jane A. Baran, and Dr. Raleigh O. Jones, are among a very small number of teams truly able to meet the challenge. They have dealt with the issues and problems surrounding the evaluation of auditory disorders for many years, devised a number of the tools in current use for clinical evaluation, and applied those tools successfully in a variety of settings. They have been in the trenches. This team also reflects a fine working relationship between audiology, otology, and neurology—critical to understanding the complex issues related to auditory disorders.

From a historical perspective, this volume might be viewed as the culmination of a steady march toward greater sophistication in the evaluation of auditory disorders. The 19th century otologists, mostly in Germany, certainly differentiated conductive loss from what was then called “perceptive” loss; however, the modern era of diagnostic auditory evaluation began in 1948 with the historic paper by the British team of M. R. Dix, C. S. Hallpike, and J. D. Hood in the *Proceed-*

ings of the Royal Society of Medicine, which demonstrated that unilateral perceptive loss (what we now call sensorineural loss) could be further differentiated into its cochlear and auditory nerve components by means of the alternate binaural loudness balance (ABLB) test.

This seminal observation stimulated international interest throughout the decades of the 1950s and 1960s in the development of a variety of methods for differentiating cochlear disorders from eighth nerve disorders, based variously on the intensity difference limen (DL), Békésy audiometric threshold tracings, nonlinear distortion of tonal stimuli, the acoustic reflex, and variations on speech audiometry. The discovery of the auditory brainstem response (ABR) in the early 1970s, however, radically altered the search. It may be difficult for young audiologists today to appreciate how this single test altered the landscape of auditory diagnostic evaluation. In addition to its value in differentiating cochlear from auditory nerve sites, it has become the sine qua non of infant hearing screening, pediatric assessment of hearing loss, and evaluation of auditory neuropathy/auditory dyssynchrony disorder. Just a few years later, the discovery of otoacoustic emissions (OAE) added another powerful

tool to the audiologist's differential diagnostic armamentarium. Today, the combination of ABR and OAE is perhaps our most powerful set of diagnostic tools.

Interest in disorders central to the auditory periphery was advanced by many audiologists, especially in the United States, following the pioneering observations of Italian investigators led by Ettore Bocca. In the mid-1950s, he and his associates demonstrated that "sensitized" speech audiometric measures could be used to reflect disorders at the level of the temporal lobes. Early in the next decade, Canadians Brenda Milner and Doreen Kimura demonstrated the right ear advantage/left ear disadvantage in dichotic listening. These two sets of observations suggested to many audiologists that speech audiometric tests, especially the dichotic variety, might identify children and adults

with "central" auditory processing disorders. Later, frequency pattern, temporal pattern, and temporal resolution tests supplemented these measures.

The authors of the present volume continue to be particularly active in this arena. In this new edition they bring us very nicely up to date on the present status of what has become, over the last five decades, a broad array of auditory disorders. This second edition especially emphasizes more case studies, coverage of additional disorders, and some very sophisticated new illustrations. In addition, the all-important reference list has been updated.

I continue to recommend this book, especially this new second edition, as an excellent text for an advanced undergraduate or graduate course on auditory disorders and their evaluation.

—James Jerger
Lake Oswego, Oregon
August 2019

About the Authors

Frank E. Musiek, PhD, is a renowned hearing researcher, scholar, teacher, and clinical audiologist. His research on electrophysiology and central auditory processing and neuroanatomy has led to the discovery and implementation of numerous tools that are widely used for assessment of the auditory brainstem and central auditory pathways. His research career has contributed in a substantial way to our fundamental understanding of the anatomy, physiology, and neurophysiology of the human auditory system. He was the recipient of the James Jerger Career Award Research in Audiology from the American Academy of Audiology in 2007 and recipient of the American Speech-Language-Hearing Association's Honors of the Association for Audiology and Auditory Neuroscience in 2010. Dr. Musiek has published more than 170 articles in peer-reviewed journals and has authored or edited 11 books.

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Jane A. Baran, PhD, is Professor Emerita in the Department of Communication Disorders at the University of Massachusetts Amherst. She is coauthor of five books and the author or coauthor of an impressive number of journal articles and book chapters in the areas of normal and disordered auditory processing, auditory electrophysiology, and neuroanatomy and neurophysiology of the auditory system. She also has presented numerous research papers and educational workshops on these topics at regional, national, and international meetings. Dr. Baran is a Fellow of the American Speech-Language-Hearing Association (ASHA), the 2001 recipient of the Clinical Educator Award from the American Academy of Audiology, a 2005 recipient of ASHA's Recognition Award for Special Contributions to Higher Education, and the 2013 recipient of the Journal of the American Academy of Audiology Editor's Award.

Raleigh O. Jones, MD, is Professor of Surgery and Chair of the Department of Otolaryngology-Head and Neck Surgery at the University of Kentucky Medical Center. He is a graduate of the University of Kentucky College of Medicine and the otolaryngology residency program at the University of North Carolina at Chapel Hill Hospitals. He completed a fellow-

ship in neurotology at the Ear Research Foundation of Florida and has specialized in the medical and surgical treatment of patients with disorders of hearing and balance. He is a member of the Triological Society and the American Otologic Society. He has authored more than 30 articles and 8 book chapters and has been awarded several teaching awards.

Acknowledgments

Writing a book such as this one, which covers a wide range of difficult topics, is no easy task. Fortunately, I had three other authors who are also great friends and who worked diligently to see this book to its finish. Jennifer Shinn, former student and friend, did an outstanding job of leading the preparation of this second edition—researching content and contributing many of the case studies. Jenn kept us going and provided much appreciated leadership and motivation—many, many thanks. Longtime colleague and friend, Jane Baran, worked tirelessly in editing this text. Her invaluable efforts and dedication need to be recognized as without her numerous contributions, this work could not have been completed. Much appreciation to Raleigh Jones, our hard-working neurotologist, whose fine contributions regarding the medical and surgical aspects of hearing disorders are the underpinnings of this book. I am honored to work with these professionals.

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Finally, we have some personal acknowledgments as this book would not have been completed without strong support from those close to us.

A heartfelt thanks to Sheila, Erik, Amy, Emma, Anna Kate, Ella, Justin, Virginia, and Simone for their love and encouragement.—FM

To my amazing family, I am forever grateful for your patience, love, and unconditional support.—JS

A very special thanks goes out to my family, who have offered their encouragement, love, and support throughout my life and my professional career.—JB

*To my wife, Dr. Jeannie Jones, whose
professional and personal sacrifices
and unwavering support have been the
underpinning of my academic career.—RJ*

Introduction

RATIONALE FOR THIS BOOK

The study of auditory disorders is the essence of a multidisciplinary approach for learning in the content areas of audiology and otology. It has been known for some time that in order to create the optimal learning format for auditory disorders, both disciplines need to contribute to the knowledge base. More recently, however, other disciplines such as neurology, pathology, and hearing science also have made important contributions to this area of study. One of the goals in writing this book was to include insights into auditory disorders from the perspectives of these various disciplines.

When the idea for writing a book on auditory disorders was originally conceptualized for the first edition, it had been a long time since such a book had been published. It was felt that an updated publication on auditory disorders directed toward AuD students, audiologists, otolaryngology residents, and other professionals in related fields was needed. In addition, based on our own personal experiences as well as considerable input from oth-

ers in audiology and related disciplines, there appeared to be a general lack of a comprehensive and full understanding of auditory disorders on the part of many graduate students and newly trained audiologists. This appeared to be especially true for disorders that affected the central auditory nervous system (CANS). This seemed perplexing because clinical audiologists are involved in the diagnosis and treatment of individuals with various disorders of the auditory system. As audiologists and health-care professionals, we play an important role in guiding patients with hearing and balance disorders through the maze of professionals who are often involved in the assessment and/or management of individuals with one or both of these particular disorders. As a result, knowledge of disorders affecting the auditory system, which is the primary focus of this text, is critical. The discussion of vestibular disorders, by design, will be limited to those disorders that present along with auditory disorders. Clinical decisions regarding whether or not to refer, to whom to refer, the timing of the referral, and why the referral is needed depend on the audiologist's knowledge of hearing and vestibular disorders, any

associated medical conditions, and the etiologic bases for these disorders and/or conditions.

The authors are now excited to publish the second edition of this book. This book covers the most common and/or significant disorders of the peripheral and central auditory systems. It has been updated to include additional disorders such as meningitis, cytomegalovirus, enlarged vestibular aqueduct syndrome, and barotrauma. Unfortunately, this book is unable to provide an exhaustive review of all auditory disorders. To stay within the scope of the book, decisions had to be made as to which disorders should be included in this text, as well as the extent of coverage of these disorders and any related conditions.

Ongoing research continues to uncover new audiologic manifestations of auditory disorders and disease, and as new editions of this book are written, those new discoveries will be included, as appropriate. The authors acknowledge that for each disorder discussed, there may be multiple presentations. We have chosen to provide case studies, which in many cases represent more commonly encountered patient presentations, evaluations, and treatments.

With respect to vestibular disorders, it was not our intent to provide a comprehensive review of these disorders, although the reader will note that some vestibular disorders are discussed. We recognize that there is a high comorbidity of auditory and vestibular involvement in a number of disorders. Information regarding vestibular involvement for auditory disorders with concurrent vestibular symptoms and system involvement will be discussed. However, it is beyond the scope of this book to address disorders that affect only the vestibular system. There-

fore, discussions of vestibular system disorders in this book are limited to disorders where the vestibular deficits are part of an auditory–vestibular disorder complex.

The references in this text are not always the original or first articles describing the disorders covered, but rather references that we believe will provide documentation on the disorders being discussed and lead the reader to more information, when desired. In many cases, recent review articles provided a framework that could be highly useful to the readers for whom this book was intended.

The coverage of auditory disorders in this book includes a review of disorders that can compromise the central auditory system as well as the peripheral auditory system. In the authors' view, the topic of central auditory system disorders frequently has been overlooked or not covered to the degree that was warranted in past publications. In this text, disorders related to the central auditory system are a key area of coverage for a number of reasons. First, there has been and continues to be a considerable amount of new pathologic information from the areas of neurology, neurotology, and neuroaudiology that impacts the diagnosis and treatment of central auditory disorders. The coverage of this topic in this text is intended to organize this information and make it readily available to the reader. For example, relatively new findings have established the role of the central auditory system in tinnitus, which is an auditory symptom that was previously believed primarily to be a manifestation of inner ear pathology (Roberts et al., 2010). In addition, it is now known that noise-induced hearing loss, as well other peripheral disorders, may have significant effects on central auditory system function and long-term

integrity (Eggermont, 2017). Therefore, an understanding of the role of the central auditory system in hearing disorders and their related symptoms, as well as the potential for central auditory system compromise secondary to peripheral involvement is important for clinicians. Second, there is both increased knowledge of and interest in central auditory disorders as documented by the sharp increase in recent publications in this area and the demand for clinical services. Therefore, it is important for audiologists and related health-care professionals to have access to information about central auditory disorders. This is needed to effectively serve both patients who are being seen for central auditory assessments, as well as individuals who may present with symptoms and preliminary audiologic findings that would result in a diagnosis of peripheral hearing loss, but who may also be at risk for central auditory compromise (Musiek, Shinn, Chermak, & Bamiou, 2017).

OVERVIEW

Chapter 2 is critical as an early chapter in this book as many clinical activities require a comprehensive understanding of auditory and vestibular anatomy and physiology. Certainly, understanding disorders of the auditory system requires such knowledge. For example, if one does not understand the underlying physiologic mechanisms associated with the inner ear and vestibular systems, one cannot fully understand or appreciate the multisystem complexities of a disorder such as Ménière's disease.

Serious discussion of auditory and vestibular disorders, both peripheral and central, requires a working knowledge

of the structure and function of the auditory and vestibular systems. This chapter focuses on the human auditory and vestibular systems to provide a reference and framework for the presentation of the hearing disorders and their sites of lesion, which are included in subsequent chapters. As mentioned previously, both peripheral and central aspects of auditory biology are overviewed; however, the reader interested in a more comprehensive discussion of the anatomy and physiology of the auditory system is referred to Musiek and Baran (2020).

Chapter 3 addresses the auditory, vestibular, and radiologic test procedures discussed in this book. Although much of the audiologic information presented may be familiar information for audiologists, it may be useful to the student or the non-audiologist reading this text. Throughout this book there has been a serious attempt to use similar forms, symbols, and terminology. A large number of case studies are included in this text, and consistency in the use of forms, symbols, and terminology should help reading efficiency. This becomes more evident when case studies utilizing vestibular, central auditory, and evoked potential assessments are reported. Chapter 3 provides an overview of the tests discussed in this text. While efforts have been made to present classic cases, it is important to note that often in clinical practice there are deviations from the "classic" presentations. This is reflected in some of the selected case studies in this text.

This chapter also includes an overview on contemporary radiology. Radiologic interpretation, of course, is based on anatomy. Therefore, we include radiologic information following the anatomy and physiology chapter. The two most common radiologic procedures, computed

tomography (CT) and magnetic resonance imaging (MRI), are reviewed. Understanding fundamental radiology on the part of the audiologist is one area that can markedly increase relevant communication with key medical personnel. A foundation in radiology can also provide insight as to the nature of a disorder and offer an excellent cross-check of audiologic test efficiency. Comparing audiologic evaluation results with radiologic evidence is one of the foundations of diagnostic audiology. Chapter 3 provides discussion of the basics of CT and MRI in a relevant, yet understandable manner. This is followed by a comparison of the two techniques revealing the advantages and limitations of each. Updated illustrations emphasizing the high utility of CT for osseous material (such as the temporal bone, ossicles, the bony cochlea, and the internal auditory canal) are provided. Other radiologic images revealing the application of MRI in viewing soft tissue structures (such as cranial nerves, brainstem, and cerebral substrate) are also included. The examples of both normal and abnormal radiologic images in this chapter (as well as subsequent chapters) provide a highly relevant learning experience for the audiologist, student, or health-care professional who has limited knowledge of or experience in interpreting the results of radiologic testing procedures.

Chapter 4 on the external and middle ear is the first that illustrates the book's organizational scheme for the four chapters that focus on a specific site of lesion (i.e., disorders of the outer and middle ear, the inner ear, the auditory nerve, and the CANS). Each of these chapters includes a review of the relevant anatomy and its associated function as well as an introduction to each of the disorders discussed in the chapter. Also included is

pertinent information on the symptoms, incidence/prevalence, etiology/pathology, site-of-lesion, audiology, medical examination, audiologic management, and medical management for the various disorders discussed. In a few instances, the disorders do not receive a full complement of information in each of the areas listed previously. This occurred because of limited information for a particular disorder, the rareness of the disorder, and/or the scope of the chapter. Typically, but not exclusively, disorders not receiving full or extensive coverage are included under "other disorders" at the end of selected chapters.

Chapter 4 covers the classic disorders affecting the outer and/or middle ear structures such as atresia, Eustachian tube dysfunction, otitis media, cholesteatoma, paraganglioma, otosclerosis, and trauma. Mentioned briefly are external otitis, exostoses, osteomas, tympanic membrane perforations, tympanosclerosis, and ossicular chain disarticulation. Also, an audiologic mechanism that has been around for some time but is often not understood by many is highlighted. It is well known that in early otitis media there is often a low-frequency tilt to the pure-tone audiogram. This is related to negative pressure increasing the stiffness of the ossicular chain. Later in the disease process, effusion may evolve, resulting in a significant "mass effect" that results in a high-frequency loss and an overall flat audiometric configuration (Jerger & Jerger, 1981). This phenomenon is discussed in greater detail within this particular chapter.

Chapter 5 discusses disorders of the inner ear. These include trauma, noise-induced hearing loss, ototoxicity, Ménière's disease, autoimmune inner ear disease, presbycusis, superior semicircular canal dehiscence, and sudden idiopathic sen-

sorineural hearing loss. Also overviewed are enlarged vestibular aqueduct syndrome, barotrauma, meningitis, cytomegalovirus, diabetes, and perilymph fistula. Recently, there have been innovations in the study of a number of cochlear disorders. For example, one of these innovations involves noise-induced hearing loss (NIHL). A better understanding of the pathologic mechanisms underlying NIHL has emerged by the discovery of free radicals and their role in NIHL (as well as other diseases) (Kopke, Coleman, Liu, Campbell, & Riffenburgh, 2002; Tieu & Campbell, 2012). High-intensity sound results in overstimulation of the inner ear structures, which leads to the production of toxic, metabolic byproducts, termed reactive oxygen species and free radicals. These free radicals result in oxidative stress on the hair cells and their related structures, and often result in damage to these inner ear structures. This new knowledge has created the potential for new approaches to the prevention and treatment of NIHL. Chemical agents that can resist the actions of free radicals have been shown to reduce the effects of NIHL. Although, to date, most of the research in this exciting area has been on animals, approaches to reduce the effects of free radicals are close to utilization with humans.

Another interesting disorder that has both audiologic and vestibular involvement is superior semicircular canal dehiscence syndrome. This is a relatively rare vestibular disorder that recently has received much attention in otology and audiology. This often is a congenital condition that may reveal itself early on or after many years. Vestibular evoked myogenic potentials are an evolving technique that can help considerably in the diagnosis of this disorder. Given the mounting interest in this disorder and its impact in

otology and audiology, its inclusion in this chapter was warranted.

Determining exactly what to include in Chapter 6 on the auditory nerve was somewhat difficult. Clearly, tumors of the eighth nerve traditionally are the key disorder to be covered. Decisions relative to what other disorders to include presented more of a challenge. Given the general awareness of and interest in auditory neuropathy spectrum disorder, it seemed reasonable to include this disorder. Several other disorders were considered, but most were extremely rare or little was known about them. The final topic included was one of interest to all of the authors; hence, it was selected for inclusion in this chapter. This disorder is commonly referred to as vascular loop syndrome.

Chapter 6 reveals some new trends in the assessment and management of patients with eighth nerve tumors that are worth mentioning here. One is the evolution of a more conservative approach to the management of eighth nerve tumors. Small tumors in older individuals are now more commonly watched and monitored rather than immediately being surgically removed. This is because some small tumors simply do not grow or grow at a very slow rate. As such, surgery may not be necessary in some cases. Another trend that is emerging, although not without some controversy, is the bypassing of ABR testing with the immediate and direct referral for MRI testing based primarily on a consideration of the patient's history and the audiogram. The final chapter on this controversial trend has not yet been written. By skipping the ABR and proceeding immediately to MRI testing, the total cost for MRI testing for patients who are considered to be at risk for eighth nerve tumors becomes exorbitant. This is because the overwhelming majority of

people with unilateral sensorineural hearing loss, which is a primary consideration for referral for MRI testing, do not have tumors. Utilization of the ABR (a much less expensive diagnostic procedure) prior to referral for radiologic assessment would markedly reduce this over-referral for MRI testing. Each of these “trends” is discussed in greater detail in the auditory nerve chapter.

Chapter 6 also relays information about another controversial topic, that of auditory neuropathy spectrum disorder. The neurologic definition of this condition may be different from the audiologic one, and this may create some difficulties in categorization. Of concern, however, is the inclusion of such disorders as hyperbilirubinemia (kernicterus at its pathological endpoint) as auditory neuropathies. Because hyperbilirubinemia does not primarily affect the auditory nerve, but rather the central auditory system and other central nervous system structures, it is not truly a peripheral neuropathy, but rather a CANS disorder. Further discussion of this controversy is included in Chapter 7.

Vascular loops are also covered in Chapter 6. Although not a popular topic with most audiologists, the authors feel it should be. Vascular loops can result in compression of the auditory and vestibular nerves in the region of the cerebellopontine angle. This often results in auditory and/or vestibular symptoms that may mimic or be misdiagnosed as another disorder (i.e., Ménière’s disease). This topic, which focuses on auditory and vestibular mechanisms, has been and continues to be of clinical interest in the otologic and neurosurgical communities, and deserves coverage in a chapter addressing inner ear disorders (Ezerarslan, Sanhal, Kurukahvecioğlu, Ataç, & Kocatürk, 2017).

Chapter 7 addresses disorders that can affect the central auditory system. These disorders are driven more by their location than by the nature of the disease. That is, many disorders may affect the CANS if they are located within that system. On the other hand, some diseases that often can affect the central auditory system may not if their pathologic action is not in the right location (i.e., within the central auditory system). This chapter highlights vascular disorders, such as stroke, involving the middle cerebral artery (MCA) and/or its branches. This vascular complex provides blood to most of the key auditory areas of the cerebrum. It is clear that disruption of the MCA complex will result in direct damage to cortical auditory structures. This in turn creates problems in higher-level hearing/cognitive processes. Despite this well-known link between anatomic damage and dysfunction, stroke patients seldom are evaluated audiotically.

Chapter 7 also brings attention to a prominent neurologic problem that also has an association with audiology, that is, Alzheimer’s disease. Individuals with Alzheimer’s disease often suffer from both peripheral and central auditory problems. Understanding the auditory factors in Alzheimer’s and other degenerative disorders can be highly useful to patients suffering from these maladies. The use of standard audiologic tests as well as central auditory procedures, including both behavioral and electrophysiologic approaches, can provide considerable information that can help guide and counsel patients with Alzheimer’s disease and their families. Indeed, it can be difficult to determine the degree of peripheral versus central auditory involvement in the patient with Alzheimer’s disease, and for some patients (e.g., those with advanced

stages of the disease), it may not be possible to obtain valid and reliable measures of auditory function. However, for many patients, some auditory testing can be completed and the use of both objective as well as subjective measures can lead to accurate diagnosis and subsequent management of the patient's auditory deficits. Deficits on the pure-tone audiogram can and should be corroborated with otoacoustic emissions (OAEs). If these individuals have valid sensitivity deficits, amplification can be a major help. Central auditory testing in this population should rely heavily on auditory evoked potentials and laterality trends on behavioral test indices.

Special attention in this chapter focuses on surgical procedures that may disrupt the CANS. There are a number of neurologic disorders that at times may require surgery. Some of these surgical procedures can result in compromise of higher auditory processes that may affect everyday communication. These patients should be evaluated prior to and after surgery to determine overall audiologic status and to inform optimal rehabilitative planning. This is a clinical population that all too often is overlooked by audiology in general.

The topics discussed in Chapter 8 include tinnitus, hyperacusis, and auditory hallucinations. These subjects could easily have been included in the context of other chapters as these symptoms/disorders are often comorbid with other audiologic and vestibular conditions. However, with the abundance of new information in these areas, it was determined that these symptoms/disorders deserved a section of their own. Tinnitus remains somewhat of a mystery in terms of a cure. However, advances in our understanding of tinnitus have changed. For example, greater

acceptance of the role of the central nervous system in tinnitus has evolved due in part to the functional imaging work of Lockwood, Salvi, and Burkard (2002). This work (as well as similar studies by others) showed activation of the auditory cortex in patients experiencing tinnitus and has provided the basis for a new approach to the study of this symptom. Although central tinnitus has long been entertained as a possibility, this concept now has gained support from many researchers who are investigating this bothersome malady.

Hyperacusis is a disorder that presents as exaggerated or inappropriate responses to sounds that are not uncomfortably loud to a typical person. It is a symptom that is often reported by patients who also report experiencing tinnitus, which has raised speculation that there may be a shared mechanism underlying the etiology and pathology of these two disorders. However, the exact mechanisms underlying hyperacusis remain unknown, but many theories abound. These theories, as well as some of the common peripheral and central conditions that may be associated with hyperacusis, are reviewed. Evaluation and management often requires a multidisciplinary team approach and can be challenging to the clinician.

Perhaps for the first time in a book such as this, the disorder of auditory hallucinations is discussed. This topic was included for several reasons. One was that the prevalence of this disorder is much greater than most people suspect. Another was that auditory hallucinations are not a problem relegated only to those with psychiatric illnesses. In many instances, auditory hallucinations may have an auditory problem (peripheral or central) as their basis. Interestingly, there is much interest