Hyperacusis and Disorders of Sound Intolerance

Clinical and Research Perspectives

Editor-in-Chief for Audiology

Brad A. Stach, PhD

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PLURAL PUBLISHING
INC.

5521 Ruffin Road
San Diego, CA 92123

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website: http://www.pluralpublishing.com

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Typeset in 11/13 Adobe Garamond by Achorn International
Printed in the United States of America by McNaughton & Gunn

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Library of Congress Cataloging-in-Publication Data

Names: Fagelson, Marc, editor. | Baguley, David (David M.), editor.
Title: Hyperacusis and disorders of sound intolerance : clinical and research perspectives / [edited by] Marc Fagelson, David M. Baguley.
Description: San Diego, CA : Plural Publishing, [2018] | Includes bibliographical references and index.
Identifiers: LCCN 2017058142| ISBN 9781944883287 (alk. paper) | ISBN 1944883282 (alk. paper)
Subjects: | MESH: Hyperacusis—diagnosis | Hyperacusis—therapy | Tinnitus—diagnosis | Tinnitus—therapy
Classification: LCC RF293.7 | NLM WV 270 | DDC 617.8/9—dc23
LC record available at https://lccn.loc.gov/2017058142

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Preface

For most of us, life is full of sound. While there are some individuals who lead a solitary, silent, or contemplative life, the more common path is replete with auditory stimulation. On an early morning walk in a North American city before conference presentations, the two editors of this book were intentionally attentive to the sounds around them, including sirens, engine sounds, phones ringing, and shouting. Many of the sound alerts in common use have been designed so as to be as compelling, and in a sense, as annoying as possible, cutting through distractions and other sounds to demand immediate and sustained attention (Patterson, 1990; Vastfjall et al., 2014).

For many years substantial scientific and clinical effort has been expended in understanding and ameliorating the impact of reduced hearing, resulting in sophisticated technologies such as hearing aids and auditory implants, advanced surgical techniques, and intensive rehabilitation strategies. Far, far less attention and scrutiny have been given to the experiences of individuals for whom the world of sound is more intense, more vivid, and perhaps perceived as more toxic than is usual. Most audiologists and otologists know of patients for whom everyday sound evokes discomfort, distress, aversion, and in some, pain.

This book seeks to explore and help the reader begin to understand those experiences. Some challenges will be encountered. The vocabulary used to describe such experiences is varied and imprecise, including decreased, reduced, or collapsed sound tolerance, and hyperacusis. Definitions of each of these terms varies; given such fundamental differences, it is not surprising that data regarding the epidemiology and natural history of hyperacusis is sparse, and inconsistent where it does exist. Some aspects of the physiological mechanisms of loudness or sound intensity perception in the auditory brain remain obscure, and there is a disconnect between the auditory neuroscience and the clinical communities that remains difficult to bridge. The experiences of individuals with reduced sound tolerance is heterogeneous, and can vary on a day to day, or hour to hour basis, and in some this is modulated by emotional and psychological state as well as the auditory environment. Tools to assess the extent and severity of loudness tolerance symptoms are crude, and in some cases may be deeply uncomfortable for the patient, as may be the case in some methods of ascertaining the threshold of loudness discomfort using sound stimulation. There is little in the way of hard evidence regarding therapy, and which interventions might be optimal for which type of patient.

All this may seem daunting, and lead one to consider that the topic of decreased sound tolerance cannot sensibly be addressed at all. However, looking at other fields would lead us to disagree. The field of pain studies was, until recently, in a similar state to that which is observed regarding hyperacusis (Chen, 2011), but systematic and focused endeavor has led to deeper understanding of mechanisms and the development of therapies that can be rigorously assessed in well-designed clinical trials. The present volume represents an attempt to take a first step on a similar journey, and it is our hope that it will inspire and provoke colleagues in both the auditory neuroscience and the clinical (especially audiology, otology,

and neurology) communities to discuss, explore, and research hyperacusis and related phenomena. Given the present state of knowledge, different authors have used different terminology, and have varied opinions, in their chapters: while we have edited with a goal of improving clarity and consistency, we have preserved some of these variations so that distinctive opinions, interpretations, and hypotheses might be heard.

Thanks are due to each of the authors for their fine contributions, and the time and effort expended on them, to Plural Publishing for agreeing to support this work, and to our families and colleagues for their support and forbearance. We would like to express our appreciation also to the many patients who have shared their experiences of hyperacusis and other phenomena of disordered loudness perception, from whom we have gleaned knowledge, experience, and inspiration.

—Marc Fagelson
David M. Baguley

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Note. DB is funded through the NIHR Biomedical Research Centre program, however, the views expressed are those of the authors and not necessarily those of the NIHR, the NHS, or the Department of Health.

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Marc and David, as well as many other professionals and the contributors to this text, continue to learn what it means for people and their families to live with hyperacusis and other forms of sound intolerance; this book is dedicated to the many individuals, patients, and providers, who sparked and continue to nurture that interest.

SECTION I

Definitions, Measurement, and Epidemiology

1

Disorders of Sound Tolerance: History and Terminology

Marc Fagelson and David M. Baguley

Introduction

Traditionally the discipline of audiology has been concerned with the challenges faced by people with reduced hearing abilities in order to alleviate the effects associated with hearing loss. While there is a multitude of such people, and the burdens they face are certainly substantial, in recent years there has been increasing awareness of a population of people who have the experience of sound being too intense for them, rather than being too quiet. In this chapter we introduce this topic, and critically examine the various definitions and models that have been proposed, and identify where present knowledge is inadequate.

The impact of hearing loss, particularly of sensory and/or neural origin on the auditory system's loudness processing, is difficult to predict, and prone to the influence of many non-auditory factors. Although hearing aid use often improves an individual's communication ability and quality of life, the lack of a straightforward relation between a patient's sensitivity and their behavior on specific audiologic tests or routine activities precludes development of interventions for individuals with hearing impairment that are consistently as effective as, for example, glasses for myopia. A similar, complex and multifactorial relation appears to exist between disorders of loudness perception, their impact, and the handicap that is associated. This chapter will focus specifically on basic elements related to disorders of sound tolerance—items such as terminology and models, both auditory-based and psychology-based, that support the terminology—that continue to vex providers, patients, and students who seek clarity when managing patients in distress.

History

The concept that human hearing varies with the sound environment and context in which the person is listening has long been accepted, the English phrase, 'I could hear a pin drop' articulating the experience of intense and active listening. The experience of hearing becoming

acute at times of emotional intensity is also known, and used extensively in horror films: for example, the trope of increasing the loudness of an enemy's footsteps as they approach along a gravel drive is widespread. The fact that an increase in the perceived intensity of the sound environment can be involuntary is less well known, however. This forms a major plot device in the novel *The Woman in White* by Wilkie Collins (1860), although the symptom is un-named. The heroine of the novel is Miss Laura Fairlie, who as the novel opens has recently been orphaned, and has inherited some money, which renders her attractive to predatory suitors, and she seeks the protection of her uncle, Mr Frederick Fairlie. Unfortunately, he is unable to support her, as he is living as a reclusive invalid, due to a number of symptoms, one of which is a decreased ability to tolerate sound. In the first conversation between niece and uncle, held at some distance across a semi-darkened room, Frederick Fairlie articulates his situation thus:

> *Pray excuse me. But could you contrive to speak in a lower key? In the wretched state of my nerves, loud sound of any kind is indescribable torture to me. You will pardon an invalid?*

Mr. Fairlie thus describes intense sound-evoked pain, leading to invalidity. While Laura eventually transcends the multiple adverse events that befall her, her uncle's situation, which is normally seen by literary critics as an abrogation of his male responsibilities, is a major setback.

Sound intolerance, such as that described by the unfortunate Mr. Fairlie, often accompanies hearing loss across patient groups of all ages, backgrounds, and auditory history, although it may also be present in individuals with hearing thresholds within the range of normal. In practice, this situation is described in terms of dynamic range, or the signal intensities that comprise the range of values spanning the patient's threshold of sensitivity to their threshold of pain or discomfort. In hearing impaired listeners, the dynamic range is reduced in one of two ways. First, in the presence of loudness recruitment thresholds of sensitivity may be elevated while thresholds of discomfort remain essentially unchanged. Alternatively, in cases of hyperacusis, the threshold of discomfort may change over time, and become abnormally low; regardless of absolute sensitivity, the patient's dynamic range would be reduced. In extreme cases, perhaps such as that described by Mr. Fairlie, the patient's dynamic range may collapse and become limited to such a narrow range of intensities that the patient develops aversions to most routine sounds and curtails life activities accordingly.

No mention is made of Mr. Fairlie having a hearing loss, and he is probably an illustration of decreased sound tolerance with normal or age-appropriate hearing thresholds. In the case of a person with hearing loss, it is valuable to consider the paradox at work, as the presence of that hearing loss should, at least on the surface, allow a person to tolerate moderate sound levels without feeling discomfort. However, many patients with hearing loss experience inordinate loudness in the presence of what should be acceptable sound levels, powerful negative emotions in the presence of sounds that most other people do not find objectionable, or sounds that cause pain in and around the ears. Patients affected by such sounds may practice avoidance strategies to minimize their discomfort; we must also consider that strategy's shortcomings.

We hope to expand upon the idea, expressed in the book's preface, that patients would benefit from greater consensus among professionals regarding terms and definitions related to unusual sound experiences. If a patient avoids situations and environments

because of aversions to specific sounds or sound levels, then that patient's discomfort and emotional response could be exacerbated by seemingly inconsistent advice and counseling from health care providers. Accurate and effective counseling relies upon many factors; certainly, the list would include an agreed-upon lexicon. In this regard, audiologists have much to learn from the psychological and trauma literatures in which a variety of clinical approaches that target patients' beliefs and understanding take a primary role in the intervention. When basic terminology cannot be agreed upon, the potential for patient learning and adapting to a challenging condition likely decreases. In this chapter, we will review definitions and terms currently in use; our intent is to move the conversation forward in a way that prioritizes lexical items supporting the needs of patients and practitioners.

Hearing Loss and Loudness Growth

Loudness Recruitment

The development of the audiometer improved the precision with which auditory thresholds were estimated. Audiometers generated signals covering a broad range of frequency and intensity, and in addition to threshold measures facilitated identifying the relation between sound intensity and the perception of loudness. Early investigators (Fowler, 1936; Steinberg & Gardner, 1937) reported that patients with hearing loss experienced, in addition to elevated thresholds, an unusual relation between the intensity of a stimulus and the loudness it evoked. For some patients with hearing loss, a signal 10 dB SL regarding the puretone threshold (that is, signal level 10 dB higher than the patient's threshold of hearing) was rated as a loudness (i.e., on a 1 to 100 scale) higher than the value provided by a normal-hearing person. Further, the rating provided by the listener with hearing loss increased by a greater amount than that of the normal hearing listener given identical increases in stimulus level. When loudness rating was graphed as a function of stimulus level, the hearing-impaired listener functions were steeper than those for normal hearing listeners. Investigators also found that, in general, at stimulus levels higher than 90 to 95 dB SPL, regardless of the hearing loss magnitude, the loudness functions as rated by hearing-impaired listeners approached and often could be superimposed upon those functions from normal hearing listeners. This loudness growth pattern was termed "loudness recruitment" by Fowler.

The loudness recruitment label captured a few interesting aspects of auditory system function. First, Fowler and other investigators employed the term to describe the way that an array of auditory nerve fibers would be influenced by damage to the cochlear hair cells. Given that not all fibers in the auditory nerve displayed identical thresholds, the loss of hair cells should have the most substantial effect on fibers with low thresholds, or those that would under normal conditions respond to audible signals of low intensity. The change in nerve innervation would commonly follow damage to some of the cochlea's most vulnerable components, the outer hair cells. While the sensory fiber population serving the outer hair cells would not be expected to influence audibility, the weakened contribution of the outer hair cells to basilar membrane mechanics would result in reduced stimulation of inner hair cells, thereby producing mild-moderate hearing loss.

At stimulus levels exceeding 50 to 60 dB, however, the relative contribution of the outer hair cell system to cochlear mechanics