PREFACE

Management of Speech and Swallowing Disorders in Degenerative Diseases was written for clinicians who serve people with amyotrophic lateral sclerosis, Parkinson's disease, Huntington's disease, or multiple sclerosis. In most service-delivery settings, clinicians see a relatively small number of adults with degenerative neurological disease each year. Management of these populations is a challenge, however, because of their diversity and the urgency of their needs. Table P.1 provides a comparison of important features of each of the neurological diseases described in this text. As is evident in the table, each disease is unique in its neuropathology, symptoms, and medical management. In some diseases, such as amyotrophic lateral sclerosis, the rate of progression may be rapid. In others, such as Parkinson's disease, progression is slow. In multiple sclerosis, rate of progression is highly variable from person to person and from one point in time to another for one person. Typical age at onset ranges from young adulthood for multiple sclerosis to the 60s and 70s for Parkinson's disease. Degree of cognitive impairment also varies from one disease to another. Although dysarthria and dysphagia are common in all these diseases, the characteristics of the speech and swallowing disorders vary. These differences in disease features dictate differing approaches to management of speech and swallowing disorders.

We were spurred to prepare this text by our work in an outpatient setting. In 1988, we established the Neuromuscular Clinic for Speech and Swallowing Disorders through the Department of Otolaryngology, University of Washington. The goal of this clinic is to evaluate degenerative speech and swallowing disorders in their mild stages and to follow patients as progression occurs, providing intervention when appropriate. We have seen hundreds of people with degenerative neurological disease and their families. We have followed many people through a number of stages of their diseases and have been impressed by a number of aspects of this clinical work:

1. The inability to communicate (or the potential of it) and the inability to eat normally are the most distressing aspects of neurological diseases such as amyotrophic lateral sclerosis.
2. The role of the medical team in patient education is critical. Although no cure exists for any of the diseases described in this text, patients and their families need information to help them make appropriate decisions about their care.
3. An urgent need exists for appropriate staging of intervention—that is, sequencing of management so that current problems are addressed and future problems anticipated. This staging should be based on knowledge of disease progression and provision of that information to patients and their families in a timely fashion. A balance must be struck between providing intervention too early—before the individual is ready—and providing intervention too late—when there is an atmosphere of crisis or when secondary complications have arisen.
4. Finally, we have learned to appreciate that diagnosis of a degenerative disease is not synonymous with a loss of hope. The task of the medical team is to be honest, realistic, and hopeful as patients and their families cope with the day-to-day challenges they face.

TABLE P.1 Comparison of Features of Four Degenerative Diseases

	Amyotrophic lateral sclerosis	Parkinson's disease	Huntington's disease	Multiple sclerosis
Changes in neural tissues	Upper and lower motor neuron degeneration	Deficiency of dopamine, which affects basal ganglia function	Deficiency of neurotransmitter GABA in the basal ganglia	Scattered multiple lesions in the CNS
Typical symptoms	Include weakness and spasticity	Include tremor, rigidity, and bradykinesia	Include chorea, hypotonia, and rigidity	May include ataxia, tremor, weakness, and spasticity
Pharmacological intervention	No significant pharmacological intervention	Drugs are important in symptom management	Medical treatment is symptomatic	Medical treatment is generally symptomatic, but antiviral agents may affect the frequency and severity of attacks
Natural course	May be rapidly progressive	Progresses slowly	Death typically occurs 15–20 years postonset	Highly variable; most frequent pattern is remission–relapsing
Age at onset	Mean age in 50s	Usually in 60s or 70s	In midlife, usually 35–40 years	Usually in young to middle-aged adults, 18–40 years
Onset of dysarthria and dysphagia	Dysarthria and dysphagia may be initial symptoms	Dysarthria and dysphagia usually appear later in the disease	Onset of dysarthria and dysphagia varies considerably	Not universal, depends on site of lesions
Type of dysarthria	Mixed spastic–flaccid	Hypokinetic	Hyperkinetic	Mixed spastic–ataxic
Cognition	Cognition usually intact	Cognitive problems may be exhibited	Cognitive changes may occur early and worsen as disease progresses	Cognitive problems may be exhibited
Awareness of dysphagia	Good awareness of dysphagia	May be unaware of dysphagia	Dysphagia is not universal; patients' subjective reports are not reliable	Varies depending on sites of lesion

Note. CNS = central nervous system; GABA = gamma-aminobutyric acid.

Management of Speech and Swallowing Disorders in Degenerative Diseases was first published in 1995 and revised in 2004. This third edition provides an update on the many advances that have been made in the understanding and treatment of degenerative diseases. Each of the first four chapters of this text is devoted to one medical diagnosis: amyotrophic lateral sclerosis, Parkinson's disease, Huntington's disease, and multiple sclerosis. Each chapter is organized around a series of questions, which were chosen to reflect information that speech–language pathologists need to know in order to manage people with degenerative disorders appropriately. Some of the questions relate to the nature of the underlying problem: What changes in neural systems are associated with the disease? What symptoms are associated with the disease? and How is the diagnosis made? Other questions relate to speech and swallowing symptoms: How rapidly do speech changes occur in the disease? and What

information is obtained in a clinical examination of speech and swallowing disorders in adults with degenerative disease? For each of the diseases, we have organized the intervention into stages, such as mild dysarthria, moderate dysarthria, and severe dysarthria. Scales rating speech and swallowing function are provided, along with clinical examination forms and checklists. In this third edition, we have added two chapters with information that crosses all of the diseases. The fifth chapter describes the clinical examination of speech and swallowing. The sixth chapter, authored by Steven Bloch, PhD, describes the maintenance of conversational interaction in the face of degenerative disease. Finally, a glossary of operational definitions serves to familiarize readers with some of the medical and technical terms used in the text.

Management of Speech and Swallowing Disorders in Degenerative Diseases also contains patient information. Our clinical experience indicates that such material is critical. Patients and their families often meet four or five professionals during the course of one clinic visit, and they are provided with a considerable amount of information in a very short time. They may not retain much of this information unless they are provided with written information that they can review repeatedly and without time pressure. On the accompanying CD, we provide reproducible handouts that include brief descriptions of normal speech and swallowing, information on each of the diseases, and descriptions of the typical effects of each disease on speech and swallowing. Suggestions for improving speech and swallowing are also provided, along with drawings of the speech and swallowing mechanisms that may be helpful in patient education. A resource list containing addresses and Web sites of pertinent national organizations is also provided on the CD (see "Useful Addresses and Web Sites").

The planning and preparation of this text have spanned a number of years. Over that time, many people have been generous in their assistance and support. In particular, we wish to acknowledge the contributions of our patients and their families. They have supported our efforts, educated us, and asked some of the difficult questions that led us to write this text.