# Adult Neurogenic Language Disorders: Assessment and Treatment

## *A Comprehensive Ethnobiological Approach*

Second Edition



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# Introduction

In the first edition of *Adult Neurogenic Language Disorders: Assessment and Treatment. A Comprehensive Ethnobiological Approach* (1997), the first chapter emphasized understanding family dynamics across ethnic groups as these related to how aging adults are viewed and cared for within diverse families. From the literature, it was quite apparent that culture plays a significant role in familial responsibility for older adult care. Culture also determines how families cope and react to disabilities in older adult family members.

Because the literature also reported differences in health status for ethnic groups, the second chapter was devoted to explaining how chronic and disabling health conditions differ in prevalence and severity for some ethnic groups but not others. The third chapter sought to explain why some members of ethnic groups may be more comfortable with alternative medicine than with Western health care and what types of barriers prevent them from participating in primary and rehabilitative health services. Three additional chapters provided information on specific disabilities that impair language and cognition: dementia, traumatic brain injury, and cerebrovascular accidents. Within these chapters, issues on differences among ethnic groups for these disabilities were infused throughout to enable the clinician to understand how these disorders might differ between and within groups of diverse patients.

Other equally important issues have emerged during the 15 years between the first and now the most current edition. The second edition should be viewed as a further discussion of cultural diversity and its role in neurogenic communication disorders. The first two chapters of this book describe national legislative and demographic changes, the effects of health disparities, and the need for culturally competent speech-language pathology services for an increasingly multicultural, multilingual patient population.

It has not been the usual practice to begin a text on neurogenic language disorders with a discussion of health disparities; however, changing events that will impact the profession for decades call for attention to this area. Hence, Chapter 1 begins the inquiry into future demographic projections, specific health legislation, and pervasive health disparities, with emphasis on health conditions that cause disorders of language and cognition for persons who are at risk. The legislation for health care reform was enacted to eliminate barriers to health care experienced by many ethnic group members. Health disparities legislation created a national institute designed to conduct research on ways to bridge the gap between those who have enjoyed access to health care and those who have not. These developments, together with ongoing projections of major population changes over the next 40 years, strongly suggest that all health care providers, including the profession of speech-language pathology, invest in culturally competent service delivery to an increasingly multilingual and multicultural patient population. Recommendations for future research are included because an aggressive research agenda is needed to provide more evidence-based information to practitioners and students in training.

Hence, Chapter 1, on health disparities, lays the groundwork for the discussion on culturally competent service delivery in the following chapter. Chapter 2, written with Wilhelmina Wright-Harp, examines bias in assessment and treatment and provides recommendations for culturally competent care that are appropriate for clinicians and organizations that provide services to adults with neurogenic language impairments. Within this chapter are resources culled from related disciplines as well as from speech-language pathology.

Cultural competence is not a new issue for speech-language pathologists and audiologists. Several articles have appeared in print recently that address how clinicians should prepare for a multicultural population of patients in the health care setting (Chin, 2000), for adults with dysphagia (Riquelme, 2004), for adults with neurogenic language disorders (Payne, 2011), and for adults with hearing disorders (Scott & Jones, 2003; Wolf, 2004). These articles, taken together with policy statements on cultural competence from the American Speech-Language-Hearing Association (2004, 2005, 2011), are positive indicators that the profession has begun the process of structuring new paradigms for service delivery for an increasingly diverse population. It is in this spirit that Chapters 1 and 2 address critical issues in diversity for consideration by the academy, researchers, and practitioners in a text on four major neurogenic disorders: dementia, traumatic brain injury, aphasia, and right hemisphere disorders.

Chapter 3 reviews basic information on neural structures necessary for language and cognition and their functions within the central nervous system. This review is updated to include those structures identified in the literature as having specific roles in memory, attention, learning, and language. Current literature on the major disorders that impair language and cognition describe recent neuroimaging procedures that make earlier and more definitive diagnoses possible. Where changes in brain structure due to Alzheimer's disease could be seen only after a patient's death, newer neuroimaging techniques allow researchers to see changes in brain structure and function early in the progression of the disease. Additionally, advances in neuroimaging enable researchers to identify the interdependence of structures for language and confirm that structures like the cerebellum and the substantia innominata in the basal forebrain play more active roles in language, learning, and memory than previously thought.

Chapter 4 focuses on dementia and related disorders, including evaluation and treatment. The most recent classifications and descriptions of dementia are discussed in this chapter as well as current trends in assessment and treatment of language and cognitive disturbances.

Chapter 5, co-authored with colleagues Wilhelmina Wright-Harp and Alaina Davis, is devoted to understanding traumatic head injury, its effects, and patient management. In this chapter, head injury prevalence across the general population will be discussed in detail; however, attention is also given to topics that are being discussed widely in the mass media: head injuries from sports and missile blast combat injuries.

Chapter 6 details causes of aphasia with emphasis on cerebrovascular accidents and other sources of focal lesions, along with aphasia identification, classifications, assessment, and treatment. Conse-

quences of right hemisphere lesions are also discussed in this chapter, as well as appropriate assessment and treatment strategies.

As the chapters unfold, it is clear that a more traditional view of the typical adult with neurogenic language disorders will need to be rethought as part of an ongoing research agenda. In some instances, some perspectives about patients with neurogenic disorders may need to be abandoned altogether if the profession is to be ready to document evidence-based assessment and intervention strategies that are appropriate for a diverse population.

In service delivery, speech-language pathologists will need to be aware of the resources and policy statements provided by the American Speech-Language-Hearing Association on cultural competence. These resources are invaluable to enable clinicians to be respectful of and sensitive to different worldviews, primary languages, ethnic group identification, socioeconomic status, cultural values, and family structures. Given the nation's long history of health disparities, speech-language pathologists will also need to be cognizant of possible differences in severity of language and cognitive disorders and other functional disabilities that may accompany a lifetime of poor or no health care, poverty, and discrimination.

To that end, this book seeks to describe neurogenic language disorders not just as a distinct set of disordered behaviors, but as disorders that affect patients and their families within the larger context of significant changes in the national landscape. Hence, the major purposes of the second edition are: (1) to provide clinicians with the most current information on assessment and management of language and cognitive communication disorders in adults; (2) to alert clinicians about possibilities for variations in clinical presentations evidenced by diverse patient populations; (3) to offer alternative strategies for assessment and treatment for patients from diverse communities; and (4) to provide clinicians with the rationale and resources for culturally competent and appropriate assessment and treatment. Each chapter includes a list of study questions to provoke thinking about the topics. At the conclusion of the book, a glossary of terms is provided to further explain the nomenclature used throughout the text. Although the second edition was written for graduate students in training, it is hoped that practitioners in speech-language pathology will find this edition to be a useful reference for assessing and treating adults with neurogenic language disorders as well.



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# SECTION II

## *The Disorders*

# CHAPTER 4

# The Dementias

## DEFINITION AND PREVALENCE OF DEMENTIA

Dementia is characterized by abnormal changes in memory, behavior, language, cognition, and the ability to perform daily living tasks caused by a number of diseases and conditions (Alzheimer's Association, 2012). Dementia, often defined by criteria from the *Diagnostic and Statistical Manual of Mental Disorders*, Fourth Edition (DSM-IV; American Psychiatric Association, 2000a), includes a decline in memory and a deficit severe enough to interfere with daily life in at least one of the following cognitive abilities:

1. Ability to generate coherent speech or understand spoken or written language.
2. Ability to recognize or identify objects, assuming intact sensory function.
3. Ability to execute motor activities, assuming intact motor abilities and sensory function and comprehension of the required task.
4. Ability to think abstractly, make sound judgments, and plan and carry out complex tasks.

Dementia is a growing national problem affecting millions of persons. Recent estimates suggest that 13.9% of persons aged 71 and older in the United States have some form of dementia (Plassman et al., 2007). Although previous observations of a leveling off of incidence among the oldest old have been reported, it is anticipated that dementia incidence may continue to increase because of the aging and the expected longer life expectancies of baby boomers.

Alzheimer's disease (AD) is the most prevalent of the dementias. More than five million Americans, or one in eight older Americans, are living with AD, which is the sixth-leading cause of death in the United States and the only cause of death among the top 10 in the United States that cannot be prevented, cured, or even slowed (Alzheimer's Association, 2012).

Establishing a definitive diagnosis of dementia includes ruling out conditions that can be reversed with treatment or surgery, specifically depression, delirium, side effects from medications, metabolic conditions like thyroid problems, vitamin deficiencies, infections, intracranial masses, arteriosclerotic complications, speech and hearing disorders, epilepsy, neurosyphilis, normal pressure hydrocephalus, chemo brain, or alcohol abuse (American Psychiatric Association, 2000b; Kean & Locke, 2008; Tonkovich, 1988). By

contrast, dementia is irreversible and is caused by permanent damage to neurons and their connections.

## EARLY SIGNS OF DEMENTIA

Dementia affects the patient, the family, and community health service providers. These dynamics may intrude upon when and how an evaluation of dementia will be sought, whether treatment for dementia complications will be initiated, and whether a patient will comply with treatment. A definitive diagnosis is often not made until the person is well into the course of dementia for a variety of reasons. One possible reason is that family members, who have not been alarmed by the subtle changes in memory or reasoning they have observed for many years, may not fully understand the ramifications of dementia symptoms or what constitutes nonpathological aging changes. Many of the observed behavioral and cognitive changes of dementia are, therefore, dismissed as normal aging.

Another possibility is that the interplay of cultural dynamics, discussed in Chapter 1, may prolong the time when family members actively seek treatment because of a perceived stigma of mental illness or distrust of the health care system. Still another possible explanation is that families may not have access to routine health care or to specialists who can diagnose dementia.

Memory loss alone is an insufficient criterion for dementia; however, an early diagnosis of AD is critically important to developing appropriate interventions. Given recent innovations in the diagnosis and treatment of dementia in the areas of diagnostics using positron emission tomography PET scans with amyloid-imaging agent Pittsburgh Compound B and retinal vascular biomarkers, and therapies using deep brain stimulation and new drug regimens (Lyketos, 2012; Rabin, 2012a), there is promise for early diagnoses and treatment.

Distinguishing between mild cognitive decline and other forms of dementia may still be difficult and may require an interdisciplinary evaluation for both cognitive and functional status. Santacruz and Swagerty (2001) provide five broad categories of signs and symptoms that should prompt an evaluation for dementia, which are:

- **cognitive changes**, such as new forgetfulness and disorientation
- **psychiatric symptoms**, such as withdrawal and apathy
- **personality changes**, such as inappropriate friendliness or flirtatiousness
- **problem behaviors**, such as agitation and wandering
- **changes in day-to-day functioning**, such as difficulty driving and getting lost.

In addition to these changes and behaviors, there are several risk factors that will need to be taken into consideration when evaluating dementia.

## RISK FACTORS FOR DEMENTIA

Although many of the irreversible dementing illnesses either are rare or have no established etiology, there are several risk factors that should become part of the case history intake for a diagnosis of dementia.

## Advancing Age

The greatest risk factor for AD and other dementing illnesses or conditions is advancing age. Most people with AD are diagnosed at age 65 or older. These individuals are said to have late-onset Alzheimer's disease. However, people younger than age 65 can also develop the disease. When AD develops in a person younger than age 65, it is considered as younger-onset or early-onset Alzheimer's disease.

## Family History

Individuals who have a parent, brother, or sister with AD are more likely to develop the disease than those who do not have a first-degree relative with AD. Those who have more than one first-degree relative with Alzheimer's are at even higher risk of developing the disease. Heyman and his colleagues (1984) found that first-degree biological relatives of others with AD are more likely to develop the disease because it may be an autosomal dominant trait. In an investigation of Down syndrome patients (Yates, Simpson, Maloney, Gordon, & Reid, 1980) found that those persons who lived to middle age developed neuritic tangles and neurofibrillary tangles characteristic of AD. Although the data were inconclusive and are still under investigation, it does appear that there may be a chemical relationship between the two syndromes through the cerebrovascular amyloid protein.

Differences in thinking skills among twins may reflect a genetic risk for dementia (Gatz, Reynolds, Finkel, Pederson, & Walters, 2010). However, cognitive changes and elevated genetic risk do not always predict that twins or siblings of people with dementia will eventually develop dementia themselves. When diseases run in families, heredity (genetics), shared environmental/lifestyle factors, or both may play a role.

## Traumatic Brain Injury

Traumatic brain injury (TBI) is associated with an increased risk of AD and other dementias. Moderate head injuries are associated with twice the risk of developing AD compared with no head injuries, and severe head injuries are associated with 4.5 times the risk (Lye & Shores, 2000). Moderate head injury is differentiated from mild head injury by the loss of consciousness or posttraumatic amnesia that lasts more than 30 minutes; if either of these lasts more than 24 hours, the injury is considered severe. These increased risks have not been shown for individuals experiencing mild head injury.

Of the several studies that have been conducted (Adle-Bassette et al., 1996; Chandra, Kokmen, Schoenberg, & Beard, 1989; Heyman et al., 1984; Katzman et al., 1996; Mortimer, 1983; Plassman et al., 2007; Rudelli, Strom, & Ambler, 1982; Tang et al., 1996; Vickrey et al., 2006), the consensus is that a history of head injury, particularly with loss of consciousness, may damage the blood–brain barrier, among other structures, which allows the pathological changes of AD to occur.

Groups that experience repeated head injuries, such as boxers, football players, and combat veterans, may be at increased risk of dementia, late-life cognitive impairment, and evidence of tau tangles at autopsy. Tau is an important protein found in neurons (Roberts, Allsop, & Bruton, 1990). Others suggest that carriers

of apolipoprotein E–epsilon 4 (ApoE-e4) who experience moderate or severe head injury are at higher risk of developing AD than ApoE-e4 carriers who do not have a history of moderate or severe head injury. The ApoE gene encodes a protein that helps regulate the levels and distribution of cholesterol and other lipids in the body (Alzheimer's Association, 2012).

## Cardiovascular Disease

The health of the brain is closely linked to the overall health of the heart and blood vessels. The brain is nourished by one of the body's richest networks of blood vessels, as seen in Chapter 3. A healthy heart helps ensure that enough blood is pumped through these blood vessels to the brain, and healthy blood vessels help ensure that the brain is supplied with the oxygen- and nutrient-rich blood it needs to function normally.

Some data show that cardiovascular disease risk factors, such as physical inactivity, high cholesterol (especially in midlife), hypertension, diabetes, smoking, and obesity, are associated with a higher risk of developing AD and other dementias (Anstey, von Sanden, Salim & O'Kearney, 2007; Kramer et al., 1999; Kramer & Erickson, 2007; Lorenzen & Murray, 2008; Pendlebury & Rothwell, 2009; Rusanen, Kivipelto, Quesenberry, Zhou, & Whitmer, 2010; Whitmer et al., 2008; Yaffe et al., 2011). Small-vessel cerebrovascular disease, visualized as white matter hyperintensities or white matter lesions on magnetic resonance imaging (MRI) scans, may be a key factor independently predictive of AD. Furthermore, among persons with mild cognitive impairment, the appearance of white matter hyperintensities may be predictive also of a later diagnosis of AD (Provenzano et al., 2013). Unlike genetic risk factors, many of these cardiovascular disease risk factors can be changed or modified to decrease the likelihood of developing cardiovascular disease and, possibly, the cognitive decline associated with AD and other forms of dementia.

## Genetic Mutations

The only risk factor gene identified so far for late-onset AD is a gene that makes ApoE. Everyone has ApoE, which helps carry cholesterol in the blood. Only about 15% of people have the form that increases the risk of AD. It is likely that other genes also may increase the risk of AD or protect against AD, but they remain to be discovered. ApoE-e4 is one of three common forms (e2, e3, and e4) of the ApoE gene, which provides the blueprint for a protein that carries cholesterol in the bloodstream (Alzheimer's Association, 2012).

Everyone inherits one form of the ApoE gene from each parent. Those who inherit one ApoE-e4 gene have increased risk of developing AD and of developing it at an earlier age than those who inherit the e2 or e3 forms of the ApoE gene. Those who inherit two ApoE-e4 genes have an even higher risk. Unlike inheriting a known genetic mutation for AD, inheriting one or two copies of this form of the ApoE gene does not guarantee that an individual will develop Alzheimer's disease.

Another known cause of AD is a genetic mutation for genes that enable cells to digest unwanted proteins. This action is essential for brain cell survival. Mutations disrupt this cellular protein-recycling process, killing nerve cells.

A small percentage of AD cases, probably less than 1%, are caused by three known genetic mutations that interfere with cellular protein recycling. Persons with an inherited form of AD carry mutations in the presenilin proteins (PSEN1 and PSEN2) or the amyloid precursor protein (APP). These disease-linked mutations result in increased production of the longer form of amyloid-beta (main component of amyloid deposits found in AD brains). Presenilins are thought to regulate APP processing. Inheriting any of these genetic mutations guarantees that an individual will develop AD. Mutations in PSEN1 and PSEN2 genes account for the majority of cases of early-onset familial AD. In such individuals, the disease tends to develop before age 65, sometimes in individuals as young as age 30. There may also be a link between PSEN1 and a familiar form of frontotemporal dementia (Alzheimer's Association, 2012; Vetrivel, Zhang, Xu, & Thinakaran, 2006).

Huntington's disease (HD) is an example of a progressive neurological disorder with dementia that has a genetic profile. Huntington's disease is an autosomal dominant disease, meaning that in affected individuals, one gene of the gene pair (the HD gene) is not functioning correctly and expresses itself more strongly, or dominates, the other working gene. Since the gene pair is not on one of the sex chromosomes, the HD gene can affect both males and females, and each gender has the same chance of having affected children (Stipe et al., 1979). A variation in the genetic code for a gene on chromosome 4 was found among persons affected with HD. Normally, the genetic code for this gene has three DNA bases, CAG, which are repeated several times. In HD, the gene sequence appears as a triplet (Indiana University School of Medicine, 1995).

## Mild Cognitive Impairment

Mild cognitive impairment (MCI) is a condition in which an individual has mild but measurable changes in thinking abilities that are noticeable to the person affected and to family members and friends but does not affect the individual's ability to carry out everyday activities (Rabin, 2012b). People with MCI, especially MCI involving memory problems, are more likely to develop AD and other dementias than people without MCI. Approximately 10% to 15% of persons with MCI develop AD, and 10% to 30% of persons with MCI have less cortical gray matter compared with healthy controls in areas known to be affected by AD pathology (Prestia et al., 2010).

Petersen (2004) identified three distinct types of MCI based on specific effects on cognition and language. In his opinion, amnestic MCI (a-MCI), characterized by memory loss, is the most common type of MCI and the most likely to convert to AD. A second subtype of MCI is single nonmemory domain MCI (sd-MCI), in which the predominant features are significant deficits in language and executive and visuospatial functions. The third subtype is a multiple-domain MCI (md-MCI) that is characterized by multiple cognitive deficits and may occur with or without significant memory loss.

Mild cognitive impairment can be reversed in some patients when caused by certain medications. However, compared with normal older adults, those with MCI with memory and those with MCI with language deficits were significantly more