

Provided for non-commercial research and education use.
Not for reproduction, distribution or commercial use.



This article appeared in a journal published by Elsevier. The attached copy is furnished to the author for internal non-commercial research and education use, including for instruction at the authors institution and sharing with colleagues.

Other uses, including reproduction and distribution, or selling or licensing copies, or posting to personal, institutional or third party websites are prohibited.

In most cases authors are permitted to post their version of the article (e.g. in Word or Tex form) to their personal website or institutional repository. Authors requiring further information regarding Elsevier's archiving and manuscript policies are encouraged to visit:

<http://www.elsevier.com/copyright>



ELSEVIER

Available online at www.sciencedirect.com

Journal of Communication Disorders 43 (2010) 438–452

Journal of
**Communication
 Disorders**

Tutorial paper

Cognition, language, and clinical pathological features of non-Alzheimer's dementias: An overview

Jamie Reilly ^{a,*}, Amy D. Rodriguez ^b, Martine Lamy ^b, Jean Neils-Strunjas ^c

^a *Department of Speech, Language, and Hearing Sciences, University of Florida, P.O. Box 117420, Dauer Hall, Gainesville, FL 32610, United States*

^b *Department of Psychiatry, University of Cincinnati, Cincinnati, OH, United States*

^c *Department of Communication Sciences and Disorders, University of Cincinnati, Cincinnati, OH, United States*

Received 3 July 2009; received in revised form 4 April 2010; accepted 19 April 2010

Abstract

There are many distinct forms of dementia whose pharmacological and behavioral management differ. Differential diagnosis among the dementia variants currently relies upon a weighted combination of genetic and protein biomarkers, neuroanatomical integrity, and behavior. Diagnostic specificity is complicated by a high degree of overlap in the initial presenting symptoms across dementia subtypes. For this reason, reliable markers are of considerable diagnostic value. Communication disorders have proven to be among the strongest predictors for discriminating among dementia subtypes. As such, speech-language pathologists may be poised to make an increasingly visible contribution to dementia diagnosis and its ongoing management. The value and durability of this potential contribution, however, demands an improved discipline-wide knowledge base about the unique features associated with different dementia variants. To this end we provide an overview of cognition, language, and clinical pathological features of four of the most common non-Alzheimer's dementias: frontotemporal dementia, vascular dementia, Lewy body disease dementia, and Parkinson's disease dementia.

Learning outcomes: Readers will learn characteristics and distinguishing features of several non-Alzheimer's dementias, including Parkinson's disease dementia, frontotemporal dementia, vascular dementia, and Lewy body dementia. Readers will also learn to distinguish between several variants of frontotemporal dementia. Finally, readers will gain knowledge of the term primary progressive aphasia as it relates to the aforementioned dementia etiologies.

© 2010 Elsevier Inc. All rights reserved.

1. Introduction

The World Health Organization's International Classification of Diseases (ICD-10) describes *dementia* as a spectrum of chronic progressive disorders that produce psychosocial or occupational impairment due to the compromise of higher cortical functioning within one or more of the following domains: memory, thinking, orientation, comprehension, calculation, learning capacity, language, and judgment (1992). The American Psychiatric Association's *Diagnostic and Statistical Manual of Mental Disorders-text revised (DSM-IV-TR)* offers alternative diagnostic criteria for dementia,

* Corresponding author. Tel.: +1 352 273 6161; fax: +1 352 273 6545.
 E-mail address: jjreilly@php.ufl.edu (J. Reilly).

characterized as a significant decline in social and occupational functioning due to memory impairment with one or more of the following associated disturbances: aphasia, apraxia, agnosia, or dysexecutive disorder (2000).

Both the ICD-10 and DSM-IV-TR criteria have been criticized for lack specificity in discriminating among the many dementia variants whose pathologies, phenotypes, and clinical courses are distinct (Reisberg, 2006). Moreover, recent advances in pharmacological and behavioral management have created a compelling need for etiology-specific treatment and early diagnosis among the many dementia variants. Differential diagnosis is, however, complicated by a high degree of overlap in the initial presenting symptoms of the dementias. Specificity is further complicated by the lack of a definitive, non-surgically invasive biomarker that can confirm *in vivo* diagnosis. Current diagnostic protocols for dementia rely upon probabilistic weighting of a number of factors, including protein and genetic biomarkers, assays of metabolic functioning, neuroimaging, and behavior.

With respect to behavior, speech and language characteristics have proven to be among the most reliable behavioral markers for distinguishing among dementia variants (Cycyk & Wright, 2008; Garrard, Maloney, Hodges, & Patterson, 2005; Grossman, D'Esposito et al., 1996; Neary et al., 1998). For this reason, speech-language pathology may be poised as a discipline to play an increasingly visible role in dementia diagnosis and management. This is especially true in light of the explosive projected growth of dementia in our aging population and the pragmatic necessity for promoting deinstitutionalized care for millions of affected patients (Hebert & Brayne, 1995; Hebert, Scherr, Bienias, Bennett, & Evans, 2004; Jorm, 1991; Zaccai, McCracken, & Brayne, 2005). One prerequisite for an informed contribution to dementia is an improved discipline-wide knowledge base regarding the unique characteristics of the various dementia subtypes. Clinical academic training in dementia is not yet universally mandatory for graduate education in speech-language pathology in the United States (See American Speech-Language-Hearing Association Curriculum Guidelines, 2008), nor is such training mandated in the United Kingdom, Australia, or Canada. As a consequence, many practicing speech-language clinicians may lack competence in treating patients with dementia and counseling caregivers.

Our aim in this review is to acquaint the reader with four dementia subtypes that are likely to increasingly comprise their caseloads during the next two decades. We focus on cognition, language, and the clinical pathological features of the following non-Alzheimer's disease dementias: frontotemporal dementia (FTD), vascular dementia (VaD), Lewy body disease dementia (LBD), and Parkinson's disease dementia (PDD). Although this is by no means an exhaustive list of non-Alzheimer's dementias, these represent the four most common dementing conditions behind Alzheimer's disease.

1.1. On the relation between aphasia and dementia

Aphasia in the broadest sense of the word represents an acquired disorder of language comprehension, production, and/or symbolic knowledge (LaPointe, 2005). Given this broad definition, one might suspect that aphasia is evident across virtually all of the dementias, and indeed an inspection of formal diagnostic criteria across many of the dementias confirms this suspicion (American Psychiatric Association, 2000; McKhann et al., 1984; Neary et al., 1998; Wetterling, Kanitz, & Borgis, 1996). Despite the ubiquity of aphasia in dementia, however, its qualitative nature and severity vary substantially as functions of disease process and localization. Such heterogeneity has led to a longstanding theoretical debate reflected in the dichotomy between disorders of degraded storage versus impaired linguistic access in dementia and stroke aphasia (Rapp & Caramazza, 1993; Warrington & Shallice, 1979).

Language disturbances in dementia have been historically attributed to the degradation of stored knowledge, whereas it is commonly assumed that language deficits in stroke aphasia reflect modality-specific impairment of access to intact conceptual knowledge (Adlam et al. 2006; Adlam, Bozeat, Arnold, Watson, & Hodges, 2006; Beaugard, Chertkow, Gold, & Bergman, 2001; Rogers, Ivanoiu, Patterson, & Hodges, 2006; Salmon, Butters, & Chan, 1999). Although the storage-access dichotomy provides an intuitive framework, its assumptions have met with criticism (for discussion see Rapp & Caramazza, 1993). Counterevidence for a clean dementia-stroke dissociation is derived from work demonstrating that patients with various forms of stroke aphasia (e.g., optic aphasia, transcortical sensory aphasia, and Broca's aphasia) can experience non-verbal conceptual impairments that are consistent with degraded storage accounts (Chertkow, Bub, Deaudon, & Whitehead, 1997; Corbett et al., 2009; Jefferies & Lambon Ralph, 2006; Jefferies, Patterson, & Lambon Ralph, 2008). An analogous body of research has demonstrated that patients with Alzheimer's disease often show modality-specific advantages (e.g., pictures named better than words) that are strongly suggestive of impaired access (Bayles & Tomoeda, 1983; Bayles, Tomoeda, Kaszniak, & Trosset, 1991; Ober & Shenaut, 1999). Thus, it is distinctly possible for patients with stroke aphasia to satisfy criteria for

storage impairment and for patients with dementia to behave consistently with access impairment. Variable performance in both populations has engendered perhaps the most controversy with respect to the nature of language and non-verbal conceptual impairment in primary progressive aphasia.

1.2. Primary progressive aphasia and its relation to dementia

Mesulam (1982) proposed the term primary progressive aphasia (PPA) to describe a slowly progressive language impairment that persists for a period of at least 2 years without dementia. Further diagnostic criteria for PPA include an insidious onset and gradual worsening of language (syntax, naming, word finding, and word comprehension) in the absence of other specific causes of aphasia (e.g., stroke and malignancy). Patients with PPA may have co-morbid acalculia (i.e., inability to perform arithmetic) or ideomotor apraxia but must not manifest other amnesic, personality, or dysexecutive deficits that impair activities of daily living (Mesulam, 2001, 2003a). PPA has been described as a “language-based dementia” (Mesulam, 2003b) and as “slowly progressive aphasia without generalized dementia” (Mesulam, 1982).

Contemporary with Mesulam’s research on PPA, Warrington and colleagues independently characterized language disorders associated with primary semantic impairment (i.e., semantic dementia) (Warrington, 1975, 1981). Parallel research on PPA and semantic dementia continued until Snowden and colleagues argued that PPA and semantic dementia likely represent different stages of the same disease pathology (Snowden, Goulding, & Neary, 1989; Snowden & Neary, 1993). In many circles, semantic dementia is now used synonymously with fluent PPA (see Adlam, Patterson, et al., 2006; Amici, Gorno-Tempini, Ogar, Dronkers, & Miller, 2006). However, others have cautioned for a clear distinction between semantic dementia and semantic variant PPA (Mesulam et al., 2009).

Perhaps the most compelling argument against interchangeable use of the terms PPA and semantic dementia lies in their differing levels of specificity. PPA represents a behavioral manifestation (i.e., a phenotype) of a number of possible disease etiologies, including frontotemporal dementia, Alzheimer’s disease, Parkinson’s disease, and Creutzfeldt–Jacob disease (Amici et al., 2006; Grossman & Ash, 2004; Mesulam, 2007; Mesulam et al., 2009). In contrast, semantic dementia is associated with a specific disease entity (i.e., frontotemporal lobar degeneration). Thus, it is exceedingly rare for Alzheimer’s disease to be described as semantic dementia. Yet it is not uncommon for Alzheimer’s disease to present as PPA. In fact, one recent postmortem confirmation study revealed that the distribution of disease pathology in one PPA cohort ($N = 38$) consisted of almost 1/3 Alzheimer’s disease (32%) (Knibb, Xuereb, Patterson, & Hodges, 2006). Here we have attempted to avoid conflating phenotypic nomenclature (e.g., PPA) with clinicopathological disease classification. Instead, we focus upon clinicopathological groupings beginning with frontotemporal dementia.

2. Frontotemporal dementia

FTD is the second most common dementia behind Alzheimer’s disease diagnosed in adults below the age of 65 years and the fourth most common dementia in industrialized nations (Heutink, 2000; Johnson et al., 2005; Neary, Snowden, & Mann, 2005). Whereas age is an increasing risk factor for many other forms of dementia, this is not true of FTD, whose onset follows a roughly Gaussian (normal) distribution with a mean age of onset near 60 years and tapering frequency during earlier and later decades of life (Forman et al., 2006; Johnson et al., 2005; Ratnavalli, Brayne, Dawson, & Hodges, 2002). Symptoms of FTD typically appear about a decade earlier than AD, making age of onset a distinctive feature of the disease (Hodges, Davies, Xuereb, Kril, & Halliday, 2003; Neary et al., 2005; Rascovsky et al., 2005). Survival in FTD varies from between 2 and 8 years post-onset, although there is considerable variability in clinical course and rate of progression (Hodges et al., 2003; Neary et al., 2005).

FTD is characterized by progressive atrophy of regions of frontal and temporal cortex (Cairns et al., 2007; Johnson et al., 2005; Neary et al., 2005). In early stage FTD, patients show relatively localized or circumscribed atrophy of these brain regions, but as FTD progresses, gyral atrophy is evident across both cerebral hemispheres (Chao et al., 2007; Whitwell, Anderson, Scahill, Rossor, & Fox, 2004). While the etiology of FTD is idiopathic or unknown, a genetic component linked to chromosome-17 is apparent in 40% of familial cases (Miller, 2007; Neary et al., 2005). In these cases, a mutation on an arm of chromosome-17 affects a gene known as progranulin (PGRN) whose disruption produces pathogenic encoding and aggregation of the proteins, tau and ubiquitin (Baker et al., 2006). In healthy neurons, tau plays a critical role in cytoskeletal support through the maintenance of structural integrity (Avila, Lim, Moreno, Belmonte, & Cuello, 2002). However, abnormal aggregations of these proteins destroy neurons. Recent

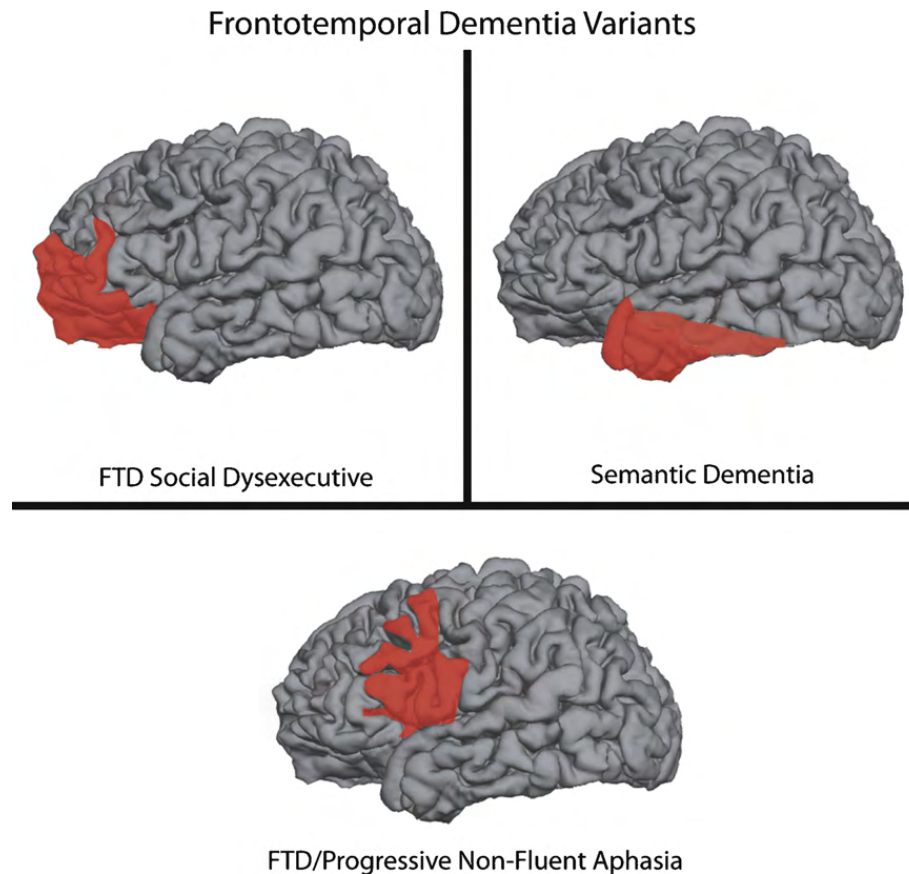


Fig. 1. Cortical atrophy in three frontotemporal dementia variants.

histopathological confirmation studies suggest that other neurodegenerative diseases such as corticobasal degeneration (CBD), Parkinsonism, progressive supranuclear palsy (PSP), motor neuron disease (MND) and amyotrophic lateral sclerosis (ALS) may all share a common protein pathology (i.e., TDP-43) (Bian & Grossman, 2007; Goldman et al., 2004; Miller, 2007).

There are at least three distinct FTD phenotypes, each having a particular distribution of cortical damage. Fig. 1 represents a schematic depiction of the following syndromes: (a) progressive non-fluent aphasia, (b) semantic dementia, and (c) frontal variant FTD (Neary et al., 1998). Common to all of these FTD subtypes is insidious onset and gradual progression in the absence of an acute precipitating event (e.g., stroke). Supportive features include an onset before the age of 65, a positive family history for FTD in a first degree relative, and the presence of bulbar palsy (motor neuron disease) (Neary et al., 1998).

2.1. Progressive non-fluent aphasia

The core diagnostic feature of PNFA outlined by Neary et al. (1998) is non-fluent speech with one or more of the following: agrammatism, phonemic paraphasias, and anomia. Supportive diagnostic features include: stuttering, oral apraxia, impaired repetition, alexia, agraphia, early preservation of word meaning, late mutism. These impairments typically occur in the context of relatively preserved episodic memory, visuospatial functioning, and temporal orientation (McKinnon et al., 2008; Peelle & Grossman, 2008; Rogers et al., 2006).

PNFA is characterized by asymmetric damage to left posterior inferior frontal cortex, including the anterior insula, basal ganglia, left inferior and middle frontal gyri, dorsal, premotor and supplementary motor cortices (Ogar, Dronkers, Brambati, Miller, & Gorno-Tempini, 2007). As left hemisphere perisylvian damage worsens, patients show increasing motor speech difficulties and impairment in phonological assembly (Croot, Patterson, & Hodges, 1998; Gorno-Tempini et al., 2006; Nestor et al., 2003). It is not uncommon for these profound difficulties in speech production to evolve to complete mutism (Gorno-Tempini et al., 2006; Neary et al., 1998). This classic

progression toward mutism in PNFA has clear implications for guiding speech-language intervention. That is, given the typical progression toward a profound deficit in speech production, one logical means of facilitating communication is through augmentative communication devices (e.g., text- or picture-to-speech). Thus, one logical strategy for PNFA may involve training patients at early stages of the disease on communication devices that will later become essential.

The distribution of left hemisphere damage in PNFA corresponds to that commonly seen in non-fluent stroke aphasia (e.g., Broca's aphasia). Thus, one might expect to see behavioral parallels between PNFA and stroke aphasia, and to some extent this is the case. Like Broca's aphasia, patients with PNFA tend to produce speech that is slow, effortful, and rife with phonemic paraphasias. Patients with PNFA have also shown disproportionate impairment for verbs relative to nouns (Hillis, Tuffiash, & Caramazza, 2002; Hillis, Sangjin, & Ken, 2004), reduced phrase length during production, and difficulties comprehending grammatically complex utterances (Ash et al., 2004; Ash et al., 2006; Grossman, D'Esposito et al., 1996; Grossman, Mickanin, et al., 1996; Peelle, Cooke, Moore, Vesely, & Grossman, 2007; Thompson, Ballard, Tait, Weintraub, & Mesulam, 1997). One recent direct comparison between non-fluent stroke aphasia and PNFA, however, revealed that the extent of syntactic loss in production (i.e., agrammatism) was worse in the stroke aphasia group relative to PNFA (Patterson, Lambon Ralph, et al., 2006). Although these differences may reflect a more posterior distribution of frontal lobe damage in PNFA, this speculative hypothesis awaits anatomical correlation.

2.2. Semantic dementia

Elizabeth Warrington reported a groundbreaking case study of patients who experienced what she described as the "selective impairment of semantic memory" (Warrington, 1975). Snowden et al. (1989) later formally classified such patients as having a distinctive neurological disorder known as *semantic dementia* (SD). On structural imaging (e.g., MRI, CT), patients with SD tend to show circumscribed atrophy affecting anterior, lateral, and ventral temporal lobe structures with relative sparing of the medial temporal lobe (Neary et al., 2005; Snowden et al., 1989). A commonly held assumption, reflected in formal diagnostic criteria, holds that there is hemispheric asymmetry with the left hemisphere more prominently affected than the right (Galton et al., 2001; Hodges, Patterson, Oxbury, & Funnell, 1992; Mummery et al., 2000; Neary et al., 1998; Snowden et al., 1989). There is, however, a growing body of literature to suggest that homologous regions of the right anterior temporal lobe are also affected (for a computational model of the spread of atrophy see Lambon Ralph, McClelland, Patterson, Galton, & Hodges, 2001).¹ Fig. 2 shows coronal, axial, and sagittal T1-weighted MR images of a patient with early to moderate semantic dementia, illustrating this typical pattern of asymmetric left anterior temporal lobe atrophy on structural imaging.

Damage to the anterior and inferolateral temporal cortex in SD produces a distinctive language profile. Patients with SD produce fluent but empty speech that is rife with deictic expressions (e.g., I don't know... It's that thing) and semantic paraphasias (e.g., 'cat' when the target word is Chihuahua) (Hodges et al., 1992; Snowden et al., 1989). These difficulties in narrative production are often amplified at the single word level and manifest as profound anomia (Lambon Ralph & Patterson, 2008; Lambon Ralph et al., 2001; Neary et al., 1998). Despite a severe naming impairment, however, patients with SD often produce over-learned phrases effortlessly. As a result, in cursory conversation it is often difficult to discern any hint of impairment. This was the first author's impression upon testing his first SD patient. His combination of fluent speech, appropriate affect, and insight masked a conceptual impairment that soon became apparent through a combination of formal testing and observation of numerous functional errors in activities of daily living (e.g., adding salt to sweeten coffee, requesting (and succeeding) in having a young investigator eat an inedible plant from his herb garden) (for a description see patient BE reported in Reilly, Martin, & Grossman, 2005).

Loss of meaning is a core diagnostic feature of SD that affects both language production and comprehension. Other core features of SD include empty spontaneous speech, semantic paraphasias, and a perceptual disorder characterized by prosopagnosia (impaired face recognition) and/or associative visual agnosia (impaired recognition of object identity) (Neary et al., 1998). For a diagnosis of SD, patients must also show relatively preserved perceptual matching

¹ The nature of hemispheric asymmetry in the progression of FTD remains controversial. Josephs et al. (2009) recently reported an imaging-behavioral correlation of the right temporal lobe variant of FTD, identifying two distinct right hemisphere FTD variants.

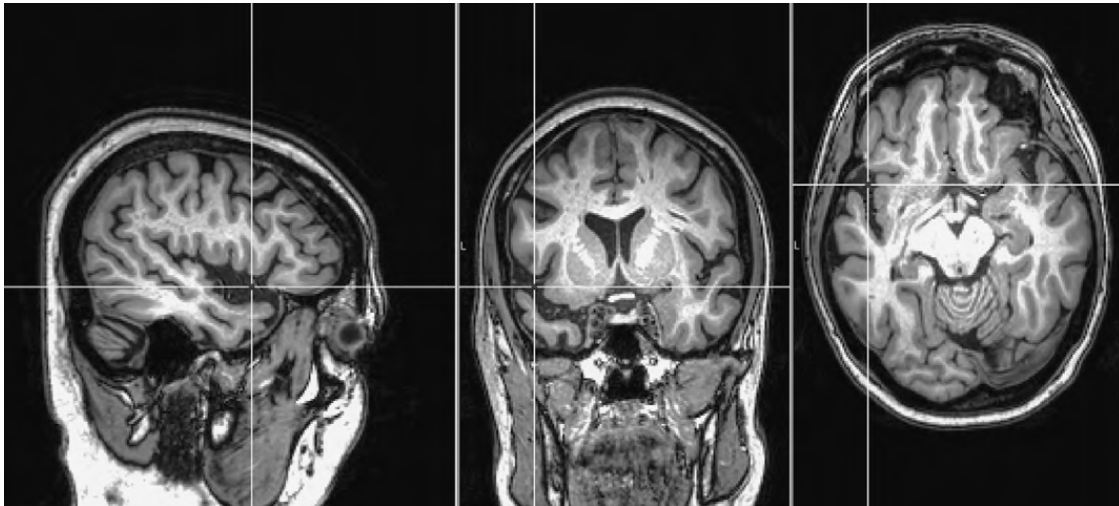


Fig. 2. Structural magnetic resonance image of semantic dementia. *Note:* Structural image of a patient with early stage semantic dementia (<12 months post-diagnosis). Asymmetric left hemisphere atrophy is evident in anterior temporal cortex as illustrated by the center of the white crosshairs.

abilities (e.g., pure tones, pictures of different people, and non-sense objects), intact single word repetition, and an ability to read aloud orthographically regular words (Neary et al., 1998).

The inability to read irregular words aloud (i.e., surface dyslexia) is ubiquitous in SD (Blazely, Coltheart, & Casey, 2005; Fushimi et al., 2003; Jefferies, Lambon Ralph, Jones, Bateman, & Patterson, 2004; Wilson et al., 2009). Surface dyslexia is a reading disorder that is characterized by regularization errors (e.g., ‘yacht’ is written to dictation as *yot* or pronounced aloud as *yaughched*) (Neary et al., 1998; Woollams, Ralph, Plaut, & Patterson, 2007). One hypothesis regarding the high frequency of surface dyslexia in SD is that patients over-rely on reading via a direct grapheme-to-phoneme conversion process (Plaut, McClelland, Seidenberg, & Patterson, 1996). That is, patients exploit their preserved phonological and orthographic abilities to compensate for degraded semantic knowledge that would normally support reading or writing (Shallice, Warrington, & McCarthy, 1983, for a more comprehensive discussion of the relative merits and disadvantages of dual route models of reading and patient-based findings see Woolams and colleagues, 2007).

Supportive features of SD include relative preservation of calculation, syntax, day-to-day memorizing (i.e., recent episodic memory), spatial skills, perception, and phonological processing. Additional impairments that are supportive of a diagnosis of SD include idiosyncratic word usage, press of speech, late-stage loss of sympathy and empathy, narrowed preoccupations, and parsimony of interactions (e.g., seclusion, limited attempts to engage in conversation) (Neary et al., 1998).

One of persistent controversy in neuroscience over the past 20 years has been the nature of the impairment underlying SD and what it can tell us about the neural correlates of semantic memory (Caramazza & Mahon, 2003; Martin, 2007; Patterson, Nestor, & Rogers, 2007; Patterson, 2007; Reilly & Peelle, 2008). The crux of this debate centers upon whether deficits reflect a combination of agnosia and aphasia or a more central loss of conceptual knowledge. One clear prediction of a central semantic deficit is that patients should experience a pervasive multi-modal impairment (Patterson, Graham, Lambon Ralph, & Hodges, 2006; Patterson, Lambon Ralph, et al., 2006). For example, loss of the concept “DOG” should permeate all possible domains of DOGNESS, manifesting as difficulties in naming a picture of a dog, recognizing the distinctive sound of a dog’s bark, and identifying the odor of a dog. The Cambridge and Manchester research groups have elegantly (and consistently) demonstrated such a broad loss of domain-specific knowledge in SD (Bozeat, Lambon Ralph, Patterson, & Hodges, 2002; Patterson, Graham, et al., 2006; Patterson, Lambon Ralph, et al., 2006; Pulvermüller et al., 2009; Rogers & McClelland, 2004; Rogers, Lambon Ralph, Hodges, & Patterson, 2004). Nonetheless, questions persist as to modality-specific advantages (e.g., reverse concreteness effects) and the role of the anterior temporal lobes in semantic memory (Simmons and Martin, 2009; Simmons, Reddish, Bellgowan, & Martin, 2010).

A few studies have tracked the longitudinal progression of SD, and a picture of the deficits in late stages of the disease has begun to emerge. Typical of this progression, Jefferies, Patterson, and Lambon Ralph (2006) report a case study of patient MK who remained receptive to testing, pleasant, and without personality change even at a very

advanced stage. For the patients we have observed, speech perception has remained remarkably intact, and we do not see the degree of speech struggle that is evident in PNFA (Kwok, Reilly, & Grossman, 2006; Reilly, Cross, Troiani, & Grossman, 2007). Nonetheless, patients do experience profound communicative impairment, and these difficulties often extend to fundamental interactions with the world.

2.3. Frontal variant FTD/behavioral variant FTD

Frontal variant FTD (FvFTD) is behaviorally distinctive from PNFA and SD (Grossman et al., 2007; Liu et al., 2004; Rascovsky et al., 2007). In FvFTD, cortical atrophy affects inferior, anterior regions of the frontal lobe (i.e., orbitofrontal cortex) (Liu et al., 2004; Rascovsky et al., 2007). Unlike PNFA and SD, individuals with FvFTD exhibit early changes in personality, organization, and attention (Neary et al., 1998, 2005). Core diagnostic criteria identified for FvFTD include a gradual decline in social conduct (i.e., inappropriate jokes, risk taking, and hypersexuality), with loss of insight and early emotional blunting (Neary et al., 1998). Supportive diagnostic features include a decline in personal hygiene, increased rigidity, hyperorality, diet changes, and the emergence of utilization behavior (i.e., compulsive use of implements such as combs or pencils that are in view of the patient).

Of the three primary FTD subtypes, FvFTD is perhaps the least likely to be referred for a speech-language clinical evaluation because social, behavioral, and emotional deficits predominate. Although it is true that aphasia is not a frank symptom of FvFTD, this disease variant does compromise language abilities. FvFTD produces impairments in executive functioning that are characterized by diminished attentional vigilance, poor inhibitory control (i.e., attending to salient stimuli while ignoring non-salient distracters), poor sequencing ability, and limitations in working memory (Rascovsky et al., 2007). In the production of narrative discourse, the net effect of these deficits is reduced thematic cohesion and disjointed event organization (Ash et al., 2006; Cooke et al., 2003; Grossman, 2002).

3. Vascular dementia

Vascular dementia (VaD), the second most frequent dementia subtype, describes a heterogeneous group of dementias associated with cerebrovascular damage (Fratiglioni et al., 2000; Traykov et al., 1999). VaD can evolve within the context of multiple cortical or subcortical strokes, cerebral hypoxemia, aneurysm, small vessel ischemic disease and genetic cerebrovascular disease. Since brain tissue requires diffuse perfusion, vascular compromise can potentially occur across many locations. Thus, the cumulative pattern of cerebrovascular damage in VaD dictates its particular phenotype. Phenotypic heterogeneity is further heightened by the fact almost half of all VaD patients have mixed vascular and Alzheimer's disease pathologies (Rocca & Kokmen, 1999; Rockwood, Bowler, Erkinjuntti, Hachinski, & Wallin, 1999; Rockwood, 1997; Traykov et al., 1999). Due to the multiplicity of potential causes of VaD, the ICD-10 has identified the following VaD variants: (1) VaD acute onset; (2) multi-infarct dementia; (3) subcortical vascular dementia; (4) mixed subcortical and cortical vascular dementia; (5) other VaD; (6) VaD unspecified (World Health Organization, 1992).

There currently exist five disparate sets of clinical criteria for VaD. These include the DSM-IV-TR (2000), the State of California Alzheimer's Disease Diagnostic and Treatment Centers (Chui et al., 1992), the National Institute of Neurological Disorders and Stroke-Association Internationale pour la Recherche et l'Enseignement en Neurosciences (NINDS-AIREN) (Roman et al., 1993), the Hachinski Ischemic Score (Hachinski et al., 1975), and the ICD-10 (1992). Postmortem clinicopathological comparisons demonstrate poor agreement amongst these criteria (Wetterling et al., 1996). It has been argued, however, that the DSM-IV-TR criteria for VaD may offer the best balance between specificity and sensitivity (Chui et al., 2000; Gold et al., 2002).

The DSM-IV-TR criteria for VaD includes social or occupational functional decline due to multiple cognitive deficits. Patients must have memory impairment and one or more of the following additional disorders: aphasia, agnosia, dysexecutive disorder. Patients must also show a combination of the following signs and symptoms in the absence of delirium: gait abnormalities, weakness of an extremity, pseudobulbar palsy, exaggeration of reflexes, and extensor plantar response. Signs must also be present in the context of laboratory evidence (e.g., structural magnetic resonance imaging) demonstrating cerebrovascular disease. Importantly, symptoms must also be judged as etiologically related to the cognitive disturbance.

Common across many reported investigations of VaD is psychomotor slowing, attentional deficits and frontal-executive dysfunction resulting in decreased self-regulation (Almkvist, Fratiglioni, Agüero-Torres, Viitanen, &

Backman, 1999; Matsuda, Saito, & Sugishita, 1998). It is also common for patients with VaD to exhibit deficits in working memory, procedural memory (e.g., tying one's shoe or using a bank machine) and episodic memory retrieval (Bowler et al., 1997; Looi & Sachdev, 1999; Reed, Eberling, Mungas, Weiner, & Jagust, 2000). Patients with VaD are likely to show decreased phonemic fluency, diminished grammatical complexity in sentence production, and flattening of pitch and amplitude contours in spontaneous speech (i.e., dysprosodia) (Jones, Laukka, & Backman, 2006; Looi & Sachdev, 1999; Lukatela, Malloy, Jenkins, & Cohen, 1998; Tierney et al., 2001). Progression of VaD is often characterized by a stepwise decline. That is, during the course of the disease, patients may experience periods of stability followed by punctuated declines in memory and other cognitive functions (Chui et al., 2000).

4. Lewy body dementia

Lewy body dementia (LBD) is a relatively new dementia classification with about one-half the frequency of AD (Rahkonen et al., 2003). Prevalence estimates for LBD vary from between 15% and 35% of all dementia cases (Geser, Wenning, Poewe, & McKeith, 2005; Zaccai et al., 2005). If the upper range of LBD (35%) is correct, then LBD represents the second most common form of dementia behind AD, relegating VaD to the third most common form of dementia (Geser et al., 2005; Heidebrink, 2002; Rahkonen et al., 2003; Weisman & McKeith, 2007; Zaccai et al., 2005). Age of onset in LBD is similar to AD, with a mean of 68 years (range 50–85 years) (Jellinger, Wenning, & Seppi, 2007; Korczyn & Reichmann, 2006; Neef & Walling, 2006). Reports on survival duration post-onset of LBD vary, with a mean of approximately 6 years (range 1.8–9.5 years) (Walker, Allen, Shergill, Mullan, & Katona, 2000).

Standard diagnosis of LBD is based on presence of dementia as well as two of the following three core diagnostic features: (1) fluctuating cognition, (2) visual hallucinations, and (3) movement disorder (McKeith, 2006). It is estimated that 50–75% of individuals with LBD experience fluctuations in cognition that range from minutes to days (McKeith et al., 2004) and 70% of individuals will develop akinetic-rigid syndrome (Geser et al., 2005). Suggestive diagnostic features include rapid eye movement sleep disorder, neuroleptic sensitivity, and low dopaminergic uptake in the basal ganglia (Galvin et al., 2008). Supportive features include syncope, postural instability with repeated falls, transient loss of consciousness, delusions, and multi-modal hallucinations (i.e., auditory and tactile) (McKeith, 2006; Weisman & McKeith, 2007).

LBD is typically associated with cognitive complaints (e.g., amnesic impairment) rather than aphasia or other language disturbance early in the course of the disease. Doubleday, Snowden, Varma, and Neary (2002) report that only 5% of LBD patients present with primary language complaints, whereas memory impairment, or a combination of memory, visuospatial, and language impairment are more commonly reported (Doubleday et al., 2002). Specific cognitive features of LBD include severe memory impairment, inattention, visuospatial disturbance, prosopagnosia, color agnosia, constructional and ideomotor apraxia, and visual distractibility (Doubleday et al., 2002; Galasko, Katzman, Salmon, & Hansen, 1996; Gibb, Luthert, Janota, & Lantos, 1989). Language disturbance in LBD is characterized by confabulation, incoherence, and perseveration during conversation, difficulty naming common objects, and a reduction in verbal fluency (Doubleday et al., 2002; Galasko et al., 1996; Gibb et al., 1989). Progression of the disease is characterized by Parkinsonism, profound memory disturbance, severe neuroleptic reactions, and disproportionate visuospatial deficits (Ferman et al., 2006; Hamilton et al., 2008). Additionally, there is some evidence that severity of visuospatial deficits predicts rate of cognitive decline (Hamilton et al., 2008). In late stages of LBD, global cognitive deficits lead to complete functional dependence.

5. Parkinson's disease dementia

Parkinson's disease (PD) has a mean age of onset of 60 years, with only 10% of those affected being 45 years of age or younger (Korchounov, Schipper, Preobrazhenskaya, Kessler, & Yakhno, 2004; Rao, Hofmann, & Shakil, 2006). The risk of developing PD increases with advanced age, and males have a greater likelihood of contracting the disease (Nussbaum & Ellis, 2003). Estimates for idiopathic PD in the world's ten most populated nations range from 4.1 to 6.6 million in 2005, with a projected expansion to 8.7–9.3 million by 2030 (Dorsey et al., 2007). In PD, dopamine depletion compromises the integrity of the cortical–striatal motor loop, producing impairment in the relative timing and initiation of purposeful movement (Louis & Frucht, 2007; Rao et al., 2006). Typically, when greater than 80% of the dopaminergic cells of the substantia nigra are depleted, individuals develop the cardinal motor symptoms of

Parkinson's disease (Louis & Frucht, 2007). These symptoms include an asymmetric onset with resting tremor, bradykinesia, rigidity, and postural instability (Jellinger et al., 2007; von Campenhausen et al., 2005).

Recent work has demonstrated that even in the absence of a frank dementia, PD is associated with a number of higher level cognitive and communication deficits (Caballol, Marti, & Tolosa, 2007; Grossman et al., 2000; Henry & Crawford, 2004; Hochstadt, Nakano, Lieberman, & Friedman, 2006; Mahieux et al., 1998; Riedel et al., 2008). Moreover, the severity of cognitive impairment in PD is strongly correlated with severity of bradykinesia, indicating a relationship between generalized cognitive and motor slowing (Levy et al., 2000; Sawamoto, Honda, Hanakawa, Fukuyama, & Shibasaki, 2002). With respect to communication, many hold that the overarching source of impairment is a dysexecutive disorder characterized by resource limitations that affect inhibitory control, speed of processing, vigilance, cognitive flexibility, and working memory (Grossman et al., 2000; Grossman, Lee, Morris, Stern, & Hurtig, 2002; Hochstadt et al., 2006). Resource-related deficits associated with prefrontal cortical dysfunction in PD produce ripple effects that are evident in tasks such as verbal fluency, sentence comprehension, and narrative production (Grossman et al., 2002; Henry & Crawford, 2004; Hochstadt et al., 2006). Interestingly, generative action naming fluency is disproportionately impaired in PD relative to semantic or phonemic fluency, and patients with PD are likely to show greater anomia for actions than objects (Cotelli et al., 2007). Other communication-related deficits include difficulties with inference, metaphor comprehension, and perception of linguistic and emotional prosody (Monetta & Pell, 2007; Pell & Leonard, 2003, 2005).

As PD progresses, as many as 70–80% of individuals convert to PD with dementia (PDD) (Emre et al., 2007). In most cases, this conversion happens within the first 10 years after onset and is insidious in nature. Upon conversion to PDD, survival duration is 5 years (mean 6.7 years), roughly similar to LBD (Emre et al., 2007; Geser et al., 2005). Predictors of survival duration in PDD and LBD include older age at onset, fluctuating cognition, severity of hallucinations, severity of akinesia, and degree of concurrent Alzheimer's disease neurofibrillary plaque and tangle pathology (Jellinger et al., 2007).

Core diagnostic criteria for PDD include an established diagnosis of PD and impairment in at least two of the following four domains: (1) attention; (2) executive functioning; (3) visuospatial processing; (4) verbal free recall. For the diagnosis of probable PDD, individuals must also show at least one behavioral symptom (i.e., apathy, depression, hallucinations, delusions, or excessive daytime sleepiness) (Emre et al., 2007).

Communication deficits include impaired concept formation, perseveration, and reduced verbal fluency across domains (Emre et al., 2007). It has also been hypothesized that reduced action word (i.e., verb) fluency predicts conversion from PD to PDD (Piatt, Fields, Paolo, Koller, & Troster, 1999). As PDD progresses, the cognitive and language deficits become more pronounced.

5.1. PDD and LBD: similarities

Histopathological investigations of both PDD and LBD reveal the presence of large, alpha-synuclein protein deposits termed Lewy bodies in the brain (Nussbaum & Ellis, 2003; Singleton et al., 2003; Theuns & Van Broeckhoven, 2008). Lewy bodies collect within the neuronal cytoplasm, ultimately destroying cells and resulting in neuronal dropout (Emre et al., 2007; Singleton et al., 2003). In PDD, Lewy body-induced cell death occurs within the substantia nigra, a midbrain structure that produces dopamine, the neurotransmitter critical for regulating communication between the basal ganglia and the motor cortex (Bear et al., 2007). In LBD, Lewy bodies are found in the substantia nigra, but are more numerous in the cerebral cortex (Byrne, Lennox, Lowe, & Godwin-Austen, 1989). In both PD and LBD, cell death is not restricted to a particular brain region. Thus, late-stage PD may resemble LBD, and LBD often evolves into a Parkinsonian syndrome (Byrne et al., 1989).

PDD and LBD have a similar set of genetic and environmental risk factors, and onset may involve an interaction of these components. In a minority of cases, PDD and LBD have been linked to several gene mutations including SNCA, PRKN, PINK1, DJ-1, and LRRK-2 (Pankratz & Foroud, 2007; Theuns & Van Broeckhoven, 2008). However, the large majority of patients with PDD or LBD have no family history of the disease and no evidence of specific gene mutations (Pankratz & Foroud, 2007). PDD and LBD share environmental factors such as exposure to pesticides, high soil concentrations of specific metals (i.e., manganese), neurotoxic drugs (i.e., synthetic heroin), and neuroleptic antipsychotic medications (Ascherio et al., 2006; Gao et al., 2007; Lai, Marion, Teschke, & Tsui, 2002).

Because of similar underlying neuropathology, there is considerable overlap between PDD and LBD with respect to clinical profiles (Ballard et al., 2006; McKeith, 2000; Neef & Walling, 2006). While LBD is typically associated with

greater memory loss and greater fluctuations in attention than PDD, no significant differences have been found between these two groups in deficits of executive function, constructional praxis, and visuospatial processing (Emre et al., 2007). This has led some to suggest that PD and LBD are spectrum disorders of Lewy body disease (Jellinger & Attems, 2008). Due to the difficulty distinguishing between LBD and PDD, current practice instantiates an arbitrary “1-year rule”, whereby dementia preceding the onset of a movement disorder by 1 year is classified as LBD, whereas dementia that occurs within the context of an existing movement disorder is classified as PDD (Emre et al., 2007).

6. Conclusion

One of the greatest public health challenges facing industrialized nations during the next three decades involves effectively coping with dementia. The psychosocial and economic necessities of promoting deinstitutionalized care for the many millions of people with dementia will soon become apparent in our rapidly aging society. As a clinical discipline, speech-language pathology may be poised to contribute valuable information through careful observation and identification of cognitive–linguistic behaviors associated with different forms of dementia. In addition, it seems equally likely that differential diagnosis will rely on identification of motor speech disorders and swallowing impairments associated with each of the subtypes. In addition, knowledge of these clinical profiles may be useful in educating patients, families and caregivers regarding the cognitive–linguistic behavioral deficits expected from onset to progression of the disease. Thus, there is great potential for speech-language pathology to emerge as a frontline discipline in dementia management. Nevertheless, it is clear that this contribution demands an improved discipline-wide knowledge base. The aim of this paper was to provide an overview of the cognitive, language and clinical pathological features of non-Alzheimer’s dementias in order to begin to address this gap in knowledge.

Acknowledgements

This work was supported by NIH/NIDCD K23 DC010197 (to JR) and the American Speech-Language-Hearing Foundation (to AR).

Appendix A. Continuing education questions

1. The “one year rule” states that a diagnosis of Parkinson’s disease dementia is appropriate when movement disorder precedes dementia by at least one year.
 - a. True
 - b. False
2. Lewy body dementia is classified as a synucleinopathy.
 - a. True
 - b. False
3. Alzheimer’s disease can produce primary progressive aphasia.
 - a. True
 - b. False
4. Progressive non-fluent aphasia and Lewy body disease are variants of frontotemporal dementia.
 - a. True
 - b. False
5. Advancing age is a risk factor for frontotemporal dementia.
 - a. True
 - b. False

References

- Adlam, A., Bozeat, S., Arnold, R., Watson, P. C., & Hodges, J. R. (2006a). Semantic knowledge in mild cognitive impairment and mild Alzheimer’s disease. *Cortex*, 42, 675–684.
- Adlam, A., Patterson, K., Rogers, T. T., Nestor, P. J., Salmon, C. H., Acosta-Cabronero, J., et al. (2006b). Semantic dementia and fluent primary progressive aphasia: Two sides of the same coin? *Brain*, 129, 3066–3080.

- Almkvist, O., Fratiglioni, L., Aguero-Torres, H., Viitanen, M., & Backman, L. (1999). Cognitive support at episodic encoding and retrieval: Similar patterns of utilization in community-based samples of Alzheimer's disease and vascular dementia patients. *Journal of Clinical Experimental Neuropsychology*, 21(6), 816–830.
- American Psychiatric Association. (2000). *Diagnostic and statistical manual of mental disorders* (4th ed.). Washington, DC: American Psychiatric Association.
- American Speech-Language-Hearing Association. (2008). *Standards for accreditation of graduate education programs in audiology and speech-language pathology*.
- Amici, S., Gorno-Tempini, M. L., Ogar, J. M., Dronkers, N. F., & Miller, B. L. (2006). An overview on primary progressive aphasia and its variants. *Behavioural Neurology*, 17(2), 77–87.
- Ascherio, A., Chen, H., Weisskopf, M. G., O'Reilly, E., McCullough, M. L., Calle, E. E., et al. (2006). Pesticide exposure and risk for Parkinson's disease. *Annals of Neurology*, 60(2), 197–203.
- Ash, S., Moore, P., Hauck, R., Antani, S., Katz, J., & Grossman, M. (2004). Quantitative analysis of paraphasic errors in frontotemporal dementia. *Neurology*, 62a, 166–183.
- Ash, S., Moore, P., Antani, S., McCawley, G., Work, M., & Grossman, M. (2006). Trying to tell a tale: Discourse impairments in progressive aphasia and frontotemporal dementia. *Neurology*, 66, 1405–1413.
- Avila, J., Lim, F., Moreno, F., Belmonte, C., & Cuellar, A. C. (2002). Tau function and dysfunction in neurons: Its role in neurodegenerative disorders. *Molecular Neurobiology*, 25(3), 213–231.
- Baker, M., Mackenzie, I. R., Pickering-Brown, S. M., Gass, J., Rademakers, R., Lindholm, C., et al. (2006). Mutations in progranulin cause tau-negative frontotemporal dementia linked to chromosome 17. *Nature*, 442(7105), 916–919.
- Ballard, C., Ziaeva, I., Perry, R., Larsen, J. P., O'Brien, J., McKeith, I., et al. (2006). Differences in neuropathologic characteristics across the Lewy body dementia spectrum. *Neurology*, 67(11), 1931–1934.
- Bayles, K. A., & Tomoeda, C. K. (1983). Confrontation naming impairment in dementia. *Brain and Language*, 19(1), 98–114.
- Bayles, K. A., Tomoeda, C. K., Kaszniak, A. W., & Trosset, M. W. (1991). Alzheimer's disease effects on semantic memory: Loss of structure or impaired processing? *Journal of Cognitive Neuroscience*, 3(2), 166–182.
- Bear, M. F., Connors, B. W., & Paradiso, M. A. (2007). *Neuroscience: Exploring the brain* (3rd ed.). Philadelphia, PA: Lippincott Williams & Wilkins Publishers.
- Beauregard, M., Chertkow, H., Gold, D., & Bergman, S. (2001). The impact of semantic impairment on word stem completion in Alzheimer's disease. *Neuropsychologia*, 39(3), 302–314.
- Bian, H., & Grossman, M. (2007). Frontotemporal lobar degeneration: Recent progress in antemortem diagnosis. *Acta Neuropathologica*, 114(1), 23–29.
- Blazely, A. M., Coltheart, M., & Casey, B. J. (2005). Semantic impairment with and without surface dyslexia: Implications for models of reading. *Cognitive Neuropsychology*, 22(6), 695–717.
- Bowler, J. V., Eliasziw, M., Steenhuis, R., Munoz, D. G., Fry, R., Merskey, H., et al. (1997). Comparative evolution of Alzheimer disease, vascular dementia, and mixed dementia. *Archives of Neurology*, 54(6), 697–703.
- Bozeat, S., Lambon Ralph, M. A., Patterson, K., & Hodges, J. R. (2002). When objects lose their meaning: What happens to their use? *Cognitive, Affective & Behavioral Neuroscience*, 2(3), 236–251.
- Byrne, E. J., Lennox, G., Lowe, J., & Godwin-Austen, R. B. (1989). Diffuse Lewy body disease: Clinical features in 15 cases. *Journal of Neurology, Neurosurgery, and Psychiatry*, 52(6), 709–717.
- Caballol, N., Marti, M. J., & Tolosa, E. (2007). Cognitive dysfunction and dementia in Parkinson disease. *Movement Disorders*, 22(S17), S358–S366.
- Cairns, N. J., Bigio, E. H., Mackenzie, I. R., Neumann, M., Lee, V. M., Hatanpaa, K. J., et al. (2007). Neuropathologic diagnostic and nosologic criteria for frontotemporal lobar degeneration: Consensus of the consortium for frontotemporal lobar degeneration. *Acta Neuropathologica*, 114(1), 5–22.
- Caramazza, A., & Mahon, B. Z. (2003). The organization of conceptual knowledge: The evidence from category-specific semantic deficits. *Trends in Cognitive Sciences*, 7(8), 354–361.
- Chao, L. L., Schuff, N., Clevenger, E. M., Mueller, S. G., Rosen, H. J., Gorno-Tempini, M. L., et al. (2007). Patterns of white matter atrophy in frontotemporal lobar degeneration. *Archives of Neurology*, 64(11), 1619–1624.
- Chertkow, H., Bub, D., Deaudo, C., & Whitehead, V. (1997). On the status of object concepts in aphasia. *Brain and Language*, 58(2), 203–232.
- Chui, H. C., Victoroff, J. I., Margolin, D., Jagust, W., Shankle, R., & Katzman, R. (1992). Criteria for the diagnosis of ischemic vascular dementia proposed by the state of California Alzheimer's disease diagnostic and treatment centers. *Neurology*, 42(3 Pt 1), 473–480.
- Chui, H. C., Mack, W., Jackson, J. E., Mungas, D., Reed, B. R., Tinklenberg, J., et al. (2000). Clinical criteria for the diagnosis of vascular dementia: A multicenter study of comparability and interrater reliability. *Archives of Neurology*, 57(2), 191–196.
- Cooke, A., DeVita, C., Gee, J., Alsop, D., Detre, J., Chen, W., et al. (2003). Neural basis for sentence comprehension deficits in frontotemporal dementia. *Brain and Language*, 85(2), 211–221.
- Corbett, F., Jefferies, E., Ehsan, S., & Lambon Ralph, M. A. (2009). Different impairments of semantic cognition in semantic dementia and semantic aphasia: Evidence from the nonverbal domain. *Brain*, 132(9), 2593–2608.
- Cotelli, M., Borroni, B., Manenti, R., Zanetti, M., Aravalo, A., Cappa, S. F., et al. (2007). Action and object naming in Parkinson's disease without dementia. *European Journal of Neurology*, 14(6), 632–637.
- Croot, K., Patterson, K., & Hodges, J. R. (1998). Single word production in nonfluent progressive aphasia. *Brain and Language*, 61(2), 226–273.
- Cyck, L. M., & Wright, H. H. (2008). Frontotemporal dementia: Its definition, differential diagnosis, and management. *Aphasiology*, 22(4), 422–444.
- Dorsey, E. R., Constantinescu, R., Thompson, J. P., Biglan, K. M., Holloway, R. G., Kiebertz, K., et al. (2007). Projected number of people with Parkinson disease in the most populous nations, 2005 through 2030. *Neurology*, 68(5), 384–386.

- Doubleday, E. K., Snowden, J. S., Varma, A. R., & Neary, D. (2002). Qualitative performance characteristics differentiate dementia with Lewy bodies and Alzheimer's disease. *Journal of Neurology, Neurosurgery, and Psychiatry*, 72(5), 602–607.
- Emre, M., Aarsland, D., Brown, R., Burn, D. J., Duyckaerts, C., Mizuno, Y., et al. (2007). Clinical diagnostic criteria for dementia associated with Parkinson's disease. *Movement Disorders*, 22(12), 1689–1707.
- Ferman, T. J., Smith, G. E., Boeve, B. F., Graff-Radford, N. R., Lucas, J. A., Knopman, D. S., et al. (2006). Neuropsychological differentiation of dementia with Lewy bodies from normal aging and Alzheimer's disease. *Clinical Neuropsychology*, 20(4), 623–636.
- Forman, M. S., Farmer, J., Johnson, J. K., Clark, C. M., Arnold, S. E., Coslett, H. B., et al. (2006). Frontotemporal Dementia: Clinicopathological Correlations. *Annals of Neurology*, 59(6), 952–962.
- Fratiglioni, L., Launer, L. J., Andersen, K., Breteler, M. M., Copeland, J. R., Dartigues, J. F., et al. (2000). Incidence of dementia and major subtypes in Europe: A collaborative study of population-based cohorts. Neurologic Diseases in the Elderly Research Group. *Neurology*, 54(11 Suppl 5), S10–S15.
- Fushimi, T., Komori, K., Ikeda, M., Patterson, K., Ijuin, M., & Tanabe, H. (2003). Surface dyslexia in a Japanese patient with semantic dementia: Evidence for similarity-based orthography-to-phonology translation. *Neuropsychologia*, 41(12), 1644–1658.
- Galasko, D., Katzman, R., Salmon, D. P., & Hansen, L. (1996). Clinical and neuropathological findings in Lewy body dementias. *Brain and Cognition*, 31(2), 166–175.
- Galton, C. J., Patterson, K., Graham, K., Lambon Ralph, M. A., Williams, G., Antoun, N., et al. (2001). Differing patterns of temporal atrophy in Alzheimer's disease and semantic dementia. *Neurology*, 57(2), 216–225.
- Galvin, J. E., Boeve, B. F., Duda, J. E., Kaufer, D., Leverenz, J. B., Lippa, C. F., et al. (2008). Current issues in Lewy body dementia diagnosis. *Treatment and Research*, 1–14.
- Gao, X., Chen, H., Fung, T. T., Logroscino, G., Schwarzschild, M. A., Hu, F. B., et al. (2007). Prospective study of dietary pattern and risk of Parkinson disease. *American Journal of Clinical Nutrition*, 86(5), 1486–1494.
- Garrard, P., Maloney, L. M., Hodges, J. R., & Patterson, K. (2005). The effects of very early Alzheimer's disease on the characteristics of writing by a renowned author. *Brain*, 128(Pt 2), 250–260.
- Geser, F., Wenning, G. K., Poewe, W., & McKeith, I. (2005). How to diagnose dementia with Lewy bodies: State of the art. *Movement Disorders*, 20(Suppl. 12), 11–20.
- Gibb, W. R., Luthert, P. J., Janota, I., & Lantos, P. L. (1989). Cortical Lewy body dementia: clinical features and classification. *Journal of Neurology, Neurosurgery, and Psychiatry*, 52(2), 185–192.
- Gold, G., Bouras, C., Canuto, A., Bergallo, M. F., Herrmann, F. R., Hof, P. R., et al. (2002). Clinicopathological Validation Study of Four Sets of Clinical Criteria for Vascular Dementia. *American Journal of Psychiatry*, 159(1), 82–87.
- Goldman, J. S., Farmer, J. M., Van Deerlin, V. M., Wilhelmsen, K. C., Miller, B. L., & Grossman, M. (2004). Frontotemporal dementia: Genetics and genetic counseling dilemmas. *Neurologist*, 10(5), 227–234.
- Gorno-Tempini, M. L., Ogar, J. M., Brambati, S. M., Wang, P., Jeong, J. H., Rankin, K. P., et al. (2006). Anatomical correlates of early mutism in progressive nonfluent aphasia. *Neurology*, 67(10), 1849–1851.
- Grossman, M. (2002). Frontotemporal dementia: A review. *Journal of the International Neuropsychological Society*, 8(4), 566–583.
- Grossman, M., & Ash, S. (2004). Primary progressive aphasia: A review. *Neurocase*, 10, 3–18.
- Grossman, M., D'Esposito, M., Hughes, E., Onishi, K., Biassou, N., White-Devine, T., et al. (1996a). Language comprehension profiles in Alzheimer's disease, multi-infarct dementia, and frontotemporal degeneration. *Neurology*, 47(1), 183–189.
- Grossman, M., Mickanin, J., Onishi, K., & Hughes, E. (1996b). Progressive nonfluent aphasia: Language, cognitive, and PET measures contrasted with probable Alzheimer's disease. *Journal of Cognitive Neuroscience*, 8(2), 135–154.
- Grossman, M., Kalmanson, J., Bernhardt, N., Morris, J., Stern, M. B., & Hurtig, H. (2000). Cognitive resource limitations during sentence comprehension in Parkinson's disease. *Brain and Language*, 73(1), 1–16.
- Grossman, M., Lee, C., Morris, J., Stern, M. B., & Hurtig, H. I. (2002). Assessing resource demands during sentence processing in Parkinson's disease. *Brain and Language*, 80(3), 603–616.
- Grossman, M., Libon, D. J., Forman, M. S., Massimo, L., Wood, E., Moore, P., et al. (2007). Distinct antemortem profiles in patients with pathologically defined frontotemporal dementia. *Archives of Neurology*, 64(11), 1601–1609.
- Hachinski, V. C., Iliff, L. D., Zilhka, E., Du Boulay, G. H., McAllister, V. L., Marshall, J., et al. (1975). Cerebral blood flow in dementia. *Archives of Neurology*, 32(9), 632–637.
- Hamilton, J. M., Salmon, D. P., Galasko, D., Raman, R., Emond, J., Hansen, L. A., et al. (2008). Visuospatial deficits predict rate of cognitive decline in autopsy-verified dementia with Lewy bodies. *Neuropsychology*, 22(6), 729–737.
- Hebert, R., & Brayne, C. (1995). Epidemiology of vascular dementia. *Neuroepidemiology*, 14(5), 240–257.
- Hebert, L. E., Scherr, P. A., Bienias, J. L., Bennett, D. A., & Evans, D. A. (2004). State-specific projections through 2025 of Alzheimer disease prevalence. *Neurology*, 62(9), 1645.
- Heidebrink, J. L. (2002). Is dementia with Lewy bodies the second most common cause of dementia? *Journal of Geriatric Psychiatry and Neurology*, 15(4), 182–187.
- Henry, J. D., & Crawford, J. R. (2004). Verbal fluency deficits in Parkinson's disease: A meta-analysis. *Journal of the International Neuropsychological Society*, 10(4), 608–622.
- Heutink, P. (2000). Untangling tau-related dementia. *Human Molecular Genetics*, 9(6), 979–986.
- Hillis, A. E., Tuffiash, E., & Caramazza, A. (2002). Modality-specific deterioration in naming verbs in nonfluent primary progressive aphasia. *Journal of Cognitive Neuroscience*, 14(7), 1099–1108.
- Hillis, A. E., Sangjin, O., & Ken, L. (2004). Deterioration of naming nouns versus verbs in Primary Progressive Aphasia. *Annals of Neurology*, 55, 268–275.
- Hochstadt, J., Nakano, H., Lieberman, P., & Friedman, J. (2006). The roles of sequencing and verbal working memory in sentence comprehension deficits in Parkinson's disease. *Brain and Language*, 97(3), 243–257.

- Hodges, J. R., Patterson, K., Oxbury, S., & Funnell, E. (1992). Semantic Dementia: Progressive fluent aphasia with temporal lobe atrophy. *Brain*, *115*(6), 1783–1806.
- Hodges, J. R., Davies, R., Xuereb, J., Kril, J., & Halliday, G. (2003). Survival in frontotemporal dementia. *Neurology*, *61*(3), 349–354.
- Jefferies, E., & Lambon Ralph, M. A. (2006). Semantic impairment in stroke aphasia versus semantic dementia: A case-series comparison. *Brain*, *129*(8), 2132–2147.
- Jefferies, E., Lambon Ralph, M. A., Jones, R., Bateman, D., & Patterson, K. (2004). Surface dyslexia in semantic dementia: A comparison of the influence of consistency and regularity. *Neurocase*, *10*(4), 290–299.
- Jefferies, E., Patterson, K., & Lambon Ralph, M. A. (2006). The Natural History of Late-stage “Pure” Semantic Dementia. *Neurocase*, *12*(1), 1–14.
- Jefferies, E., Patterson, K., & Lambon Ralph, M. A. (2008). Deficits of knowledge versus executive control in semantic cognition: Insights from cued naming. *Neuropsychologia*, *46*(2), 649–658.
- Jellinger, K. A., & Attems, J. (2008). Prevalence and impact of vascular and Alzheimer pathologies in Lewy body disease. *Acta Neuropathologica*, *114*, 427–436.
- Jellinger, K. A., Wenning, G. K., & Seppi, K. (2007). Predictors of survival in dementia with lewy bodies and Parkinson dementia. *Neurodegenerative Diseases*, *4*(6), 428–430.
- Johnson, J. K., Diehl, J., Mendez, M. F., Neuhaus, J., Shapira, J. S., Forman, M., et al. (2005). Frontotemporal lobar degeneration: Demographic characteristics of 353 patients. *Archives of Neurology*, *62*(6), 925–930.
- Jones, S., Laukka, E. J., & Backman, L. (2006). Differential verbal fluency deficits in the preclinical stages of Alzheimer’s disease and vascular dementia. *Cortex*, *42*(3), 347–355.
- Jorm, A. F. (1991). Cross-national comparisons of the occurrence of Alzheimer’s and vascular dementias. *European Archives of Psychiatry and Clinical Neuroscience*, *240*(4–5), 218–222.
- Josephs, K. A., Whitwell, J. L., Knopman, D. S., Boeve, B. F., Vemuri, P., Senjem, M. L., et al. (2009). Two distinct subtypes of right temporal variant frontotemporal dementia. *Neurology*, *73*(18), 1443–1450.
- Knibb, J. A., Xuereb, J. H., Patterson, K., & Hodges, J. R. (2006). Clinical and pathological characterization of progressive aphasia. *Annals of Neurology*, *59*(1), 156–165.
- Korchounov, A., Schipper, H. I., Preobrazhenskaya, I. S., Kessler, K. R., & Yakhno, N. N. (2004). Differences in age at onset and familial aggregation between clinical types of idiopathic Parkinson’s disease. *Mov Disord*, *19*(9), 1059–1064.
- Korczyn, A. D., & Reichmann, H. (2006). Dementia with Lewy bodies. *Journal of Neurological Sciences*, *248*(1–2), 3–8.
- Kwok, S., Reilly, J., & Grossman, M. (2006). Acoustic-phonetic processing in semantic dementia. *Brain and Language*, *99*, 145–146.
- Lai, B. C., Marion, S. A., Teschke, K., & Tsui, J. K. (2002). Occupational and environmental risk factors for Parkinson’s disease. *Parkinsonism Related Disorders*, *8*(5), 297–309.
- Lambon Ralph, M. A., & Patterson, K. (2008). Generalization and differentiation in semantic memory: Insights from semantic dementia. *Annals of the New York Academy of Science*, *1124*, 61–76.
- Lambon Ralph, M. A., McClelland, J. L., Patterson, K., Galton, C. J., & Hodges, J. R. (2001). No right to speak? The relationship between object naming and semantic impairment: Neuropsychological evidence and a computational model. *Journal of Cognitive Neuroscience*, *13*(3), 341–356.
- LaPointe, L. L. (2005). *Aphasia and related neurogenic language disorders* (3rd ed.). New York, NY: Thieme New York.
- Levy, G., Tang, M. X., Cote, L. J., Louis, E. D., Alfaró, B., Mejia, H., et al. (2000). Motor impairment in PD: Relationship to incident dementia and age. *Neurology*, *55*(4), 539–544.
- Liu, W., Miller, B. L., Kramer, J. H., Rankin, K., Wyss-Coray, C., Gearhart, R., et al. (2004). Behavioral disorders in the frontal and temporal variants of frontotemporal dementia. *Neurology*, *62*(5), 742–748.
- Looi, J. C., & Sachdev, P. S. (1999). Differentiation of vascular dementia from AD on neuropsychological tests. *Neurology*, *53*(4), 670–678.
- Louis, E. D., & Frucht, S. J. (2007). Prevalence of essential tremor in patients with Parkinson’s disease vs. Parkinson-plus syndromes. *Movement Disorders*, *22*(10), 1402–1407.
- Lukatela, K., Malloy, P., Jenkins, M., & Cohen, R. (1998). The naming deficit in early Alzheimer’s and vascular dementia. *Neuropsychology*, *12*(4), 565–572.
- Mahieux, F., Fanelon, G., Flahault, A., Manificier, M.-J., Michelet, D., & Boller, F. O. (1998). Neuropsychological prediction of dementia in Parkinson’s disease. *Journal of Neurology, Neurosurgery & Psychiatry*, *64*(2), 178–183.
- Martin, A. (2007). The representation of object concepts in the brain. *Annual Review of Psychology*, *58*, 25–45.
- Matsuda, O., Saito, M., & Sugishita, M. (1998). Cognitive deficits of mild dementia: A comparison between dementia of the Alzheimer’s type and vascular dementia. *Psychiatry Clin Neurosci*, *52*(1), 87–91.
- McKeith, I. G. (2000). Spectrum of Parkinson’s disease, Parkinson’s dementia, and Lewy body dementia. *Neurology Clinics*, *18*(4), 865–902.
- McKeith, I. G. (2006). Consensus guidelines for the clinical and pathologic diagnosis of dementia with Lewy bodies (DLB): Report of the consortium on DLB international workshop. *Journal of Alzheimer’s Disease and Associated Disorders*, *9*(3 Suppl), 417–423.
- McKeith, I., Mintzer, J., Aarsland, D., Burn, D., Chiu, H., Cohen-Mansfield, J., et al. (2004). Dementia with Lewy bodies. *Lancet Neurology*, *3*(1), 19–28.
- McKhann, G., Drachman, D., Folstein, M., Katzman, R., Price, D., & Stadlan, E. M. (1984). Clinical diagnosis of Alzheimer’s disease: Report of the NINCDS-ADRDA work group under the auspices of Department of Health and Human Services Task Force on Alzheimer’s disease. *Neurology*, *34*, 939–944.
- McKinnon, M. C., Nica, E. I., Sengdy, P., Kovacevic, N., Moscovitch, M., Freedman, M., et al. (2008). Autobiographical memory and patterns of brain atrophy in frontotemporal lobar degeneration. *Journal of Cognitive Neuroscience*, *20*(10), 1839–1853.
- Mesulam, M. M. (1982). Slowly progressive aphasia without generalized dementia. *Annals of Neurology*, *11*, 592–598.
- Mesulam, M. M. (2001). Primary progressive aphasia. *Annals of Neurology*, *49*(4), 425–432.
- Mesulam, M. M. (2003a). Primary progressive aphasia—A language-based dementia. *New England Journal of Medicine*, *349*(16), 1535–1542.

- Mesulam, M. M. (2003b). Primary progressive aphasia: A language-based dementia. *New England Journal of Medicine*, 349(16), 1535–1542.
- Mesulam, M. M. (2007). Primary progressive aphasia: A 25-year retrospective. *Alzheimer's Disease and Associated Disorders*, 21(4), S8–S11.
- Mesulam, M. M., Rogalski, E., Wieneke, C., Cobia, D., Rademaker, A., Thompson, C., et al. (2009). Neurology of anomia in the semantic variant of primary progressive aphasia. *Brain*, 132(9), 2553–2565.
- Miller, B. L. (2007). Frontotemporal dementia and semantic dementia: Anatomic variations on the same disease or distinctive entities? *Alzheimer's Disease and Associated Disorders*, 21(4), S19–S22.
- Monetta, L., & Pell, M. D. (2007). Effects of verbal working memory deficits on metaphor comprehension in patients with Parkinson's disease. *Brain and Language*, 101(1), 80–89.
- Mummery, C. J., Patterson, K., Price, C. J., Ashburner, J., Frackowiak, R. S. J., & Hodges, J. R. (2000). A voxel-based morphometry study of semantic dementia: Relationship between temporal lobe atrophy and semantic memory. *Annals of Neurology*, 47, 36–45.
- Neary, D., Snowden, J. S., Gustafson, L., Passant, U., Stuss, D., Black, S., et al. (1998). Frontotemporal lobar degeneration: A consensus on clinical diagnostic criteria. *Neurology*, 51(6), 1546–1554.
- Neary, D., Snowden, J., & Mann, D. (2005). Frontotemporal dementia. *Lancet Neurology*, 4(11), 771–780.
- Neef, D., & Walling, A. D. (2006). Dementia with Lewy bodies: An emerging disease. *American Family Physician*, 73(7), 1223–1229.
- Nestor, P. J., Graham, N. L., Fryer, T. D., Williams, G. B., Patterson, K., & Hodges, J. R. (2003). Progressive non-fluent aphasia is associated with hypometabolism centred on the left anterior insula. *Brain*, 126(Pt 11), 2406–2418.
- Nussbaum, R. L., & Ellis, C. E. (2003). Alzheimer's disease and Parkinson's disease. *New England Journal of Medicine*, 348(14), 1356–1364.
- Ober, B. A., & Shenaut, G. K. (1999). Well-organized conceptual domains in Alzheimer's disease. *Journal of the International Neuropsychological Society*, 5(7), 676–684.
- Ogar, J. M., Dronkers, N. F., Brambati, S. M., Miller, B. L., & Gorno-Tempini, M. L. (2007). Progressive nonfluent aphasia and its characteristic motor speech deficits. *Alzheimer's Disease and Associated Disorders*, 21(4), S23–S30.
- Pankratz, N., & Foroud, T. (2007). Genetics of Parkinson disease. *Genet Med*, 9(12), 801–811.
- Patterson, K. (2007). The reign of typicality in semantic memory. *Philosophical Transactions of the Royal Society of London B: Biological Sciences*, 362(1481), 813–821.
- Patterson, K., Graham, N. L., Lambon Ralph, M. A., & Hodges, J. R. (2006a). Progressive non-fluent aphasia is not a progressive form of non-fluent (post-stroke) aphasia. *Aphasiology*, 20(9–11), 1018–1034.
- Patterson, K., Lambon Ralph, M. A., Jefferies, E., Woollams, A., Jones, R., Hodges, J. R., et al. (2006b). "Presemantic" cognition in semantic dementia: Six deficits in search of an explanation. *Journal of Cognitive Neuroscience*, 18(2), 169–183.
- Patterson, K., Nestor, P. J., & Rogers, T. T. (2007). Where do you know what you know? The representation of semantic knowledge in the human brain. *Nature Reviews Neuroscience*, 8(12), 976–987.
- Peelle, J. E., & Grossman, M. (2008). Language processing in frontotemporal dementia: A brief review. *Language and Linguistics Compass*.
- Peelle, J. E., Cooke, A., Moore, P., Vesely, L., & Grossman, M. (2007). Syntactic and thematic components of sentence processing in progressive nonfluent aphasia and nonaphasic frontotemporal dementia. *Journal of Neurolinguistics*, 20(6), 482–494.
- Pell, M. D., & Leonard, C. L. (2003). Processing emotional tone from speech in Parkinson's disease: A role for the basal ganglia. *Cognitive, Affective & Behavioral Neuroscience*, 3(4), 275–288.
- Pell, M. D., & Leonard, C. L. (2005). Facial expression decoding in early Parkinson's disease. *Cognitive Brain Research*, 23(2), 327–340.
- Piatt, A. L., Fields, J. A., Paolo, A. M., Koller, W. C., & Troster, A. I. (1999). Lexical, semantic, and action verbal fluency in Parkinson's disease with and without dementia. *Journal of Clinical Experimental Neuropsychology*, 21(4), 435–443.
- Plaut, D. C., McClelland, J. L., Seidenberg, M. S., & Patterson, K. (1996). Understanding normal and impaired word reading: Computational principles in quasi-regular domains. *Psychological Review*, 103(1), 56–115.
- Pulvermüller, F., Cooper-Pye, E., Dine, C., Hauk, O., Nestor, P. J., & Patterson, K. (2009). The word processing deficit in semantic dementia: All categories are equal, but some categories are more equal than others. *Journal of Cognitive Neuroscience (Ahead of print)*.
- Rahkonen, T., Eloniemi-Sulkava, U., Rissanen, S., Vatanen, A., Viramo, P., & Sulkava, R. (2003). Dementia with Lewy bodies according to the consensus criteria in a general population aged 75 years or older. *Journal of Neurology, Neurosurgery, and Psychiatry*, 74(6), 720–724.
- Rao, S. S., Hofmann, L. A., & Shakil, A. (2006). Parkinson's disease: Diagnosis and treatment. *American Family Physician*, 74(12), 2046–2054.
- Rapp, B., & Caramazza, A. (1993). On the distinction between deficits of access and deficits of storage: A question of theory. *Cognitive Neuropsychology*, 10(2), 113–141.
- Rascovsky, K., Salmon, D. P., Lipton, A. M., Leverenz, J. B., DeCarli, C., Jagust, W. J., et al. (2005). Rate of progression differs in frontotemporal dementia and Alzheimer disease. *Neurology*, 65(3), 397–403.
- Rascovsky, K., Hodges, J. R., Kipps, C. M., Johnson, J. K., Seeley, W. W., Mendez, M. F., et al. (2007). Diagnostic criteria for the behavioral variant of frontotemporal dementia (bvFTD): Current limitations and future directions. *Alzheimer's Disease and Associated Disorders*, 21(4), S14–S18.
- Ratnavalli, E., Brayne, C., Dawson, K., & Hodges, J. R. (2002). The prevalence of frontotemporal dementia. *Neurology*, 58(11), 1615–1621.
- Reed, B. R., Eberling, J. L., Mungas, D., Weiner, M. W., & Jagust, W. J. (2000). Memory failure has different mechanisms in subcortical stroke and Alzheimer's disease. *Annals of Neurology*, 48(3), 275–284.
- Reilly, J., & Peelle, J. E. (2008). Effects of semantic impairment on language processing in semantic dementia. *Seminars in Speech and Language*, 29, 32–43.
- Reilly, J., Martin, N., & Grossman, M. (2005). Verbal learning in semantic dementia: Is repetition priming a useful strategy? *Aphasiology*, 19, 329–339.
- Reilly, J., Cross, K., Troiani, V., & Grossman, M. (2007). Single word semantic judgments in semantic dementia: Do phonology and grammatical class count? *Aphasiology*, 21(6–8), 558–569.
- Reisberg, B. (2006). Diagnostic criteria in dementia: A comparison of current criteria, research challenges, and implications for DSM-V. *Journal of Geriatric Psychiatry and Neurology*, 19(3), 137–146.

- Riedel, O., Klotsche, J., Spottke, A., Deuschl, G., Forstl, H., Henn, F., et al. (2008). Cognitive impairment in 873 patients with idiopathic Parkinson's disease: Results from the German Study on Epidemiology of Parkinson's Disease with Dementia (GEPAD). *Journal of Neurology*, 255, 255–264.
- Rocca, W. A., & Kokmen, E. (1999). Frequency and distribution of vascular dementia. *Alzheimer's Disease and Associated Disorders*, 13(Suppl. 3), S9–S14.
- Rockwood, K. (1997). Lessons from mixed dementia. *International Psychogeriatrics*, 9(3), 245–249.
- Rockwood, K., Bowler, J., Erkinjuntti, T., Hachinski, V., & Wallin, A. (1999). Subtypes of vascular dementia. *Alzheimer's Disease and Associated Disorders*, 13(Suppl. 3), S59–S65.
- Rogers, T. T., & McClelland, J. L. (2004). *Semantic cognition: A parallel distributed processing approach*. Cambridge, MA US: MIT Press.
- Rogers, T. T., Lambon Ralph, M. A., Hodges, J. R., & Patterson, K. (2004). Natural selection: The impact of semantic impairment on lexical and object decision. *Cognitive Neuropsychology*, 21(2–4), 331–352.
- Rogers, T. T., Ivanoiu, A., Patterson, K., & Hodges, J. R. (2006). Semantic memory in Alzheimer's disease and the frontotemporal dementias: A longitudinal study of 236 patients. *Neuropsychology*, 20(3), 319–335.
- Roman, G. C., Tatemichi, T. K., Erkinjuntti, T., Cummings, J. L., Masdeu, J. C., Garcia, J. H., et al. (1993). Vascular dementia: Diagnostic criteria for research studies. Report of the NINDS-AIREN international workshop. *Neurology*, 43(2), 250–260.
- Salmon, D. P., Butters, N., & Chan, A. S. (1999). The deterioration of semantic memory in Alzheimer's disease. *Canadian Journal of Experimental Psychology*, 53(1), 108–116.
- Sawamoto, N., Honda, M., Hanakawa, T., Fukuyama, H., & Shibasaki, H. (2002). Cognitive slowing in Parkinson's disease: A behavioral evaluation independent of motor slowing. *Journal of Neuroscience*, 22(12), 5198–5203.
- Shallice, T., Warrington, E. K., & McCarthy, R. A. (1983). Reading without semantics. *The Quarterly Journal of Experimental Psychology A: Human Experimental Psychology*, 35A(1), 111–138.
- Simmons, W. K., & Martin, A. (2009). The anterior temporal lobes and the functional architecture of semantic memory. *Journal of the International Neuropsychological Society*, 15(5), 645–649.
- Simmons, W. K., Reddish, M., Bellgowan, P. S., & Martin, A. (2010). The selectivity and functional connectivity of the anterior temporal lobes. *Cerebral Cortex*, 20(4), 813–825.
- Singleton, A. B., Farrer, M., Johnson, J., Singleton, A., Hague, S., Kachergus, J., et al. (2003). alpha-Synuclein locus triplication causes Parkinson's disease. *Science*, 302(5646), 841.
- Snowden, J. S., & Neary, D. (1993). Progressive language dysfunction and lobar atrophy. *Dementia*, 4(3–4), 226–231.
- Snowden, J. S., Goulding, P. J., & Neary, D. (1989). Semantic dementia: A form of circumscribed cerebral atrophy. *Behavioural Neurology*, 2, 167–182.
- Theuns, J., & Van Broeckhoven, C. (2008). alpha-Synuclein gene duplications in sporadic Parkinson disease. *Neurology*, 70(1), 7–9.
- Thompson, C. K., Ballard, K. J., Tait, M. E., Weintraub, S., & Mesulam, M. M. (1997). Patterns of language decline in non-fluent primary progressive aphasia. *Aphasiology*, 11(4–5), 297–331.
- Tierney, M. C., Black, S. E., Szalai, J. P., Snow, W. G., Fisher, R. H., Nadon, G., et al. (2001). Recognition memory and verbal fluency differentiate probable Alzheimer disease from subcortical ischemic vascular dementia. *Archives of Neurology*, 58(10), 1654–1659.
- Traykov, L., Rigaud, A. S., Caputo, L., Couderc, R., Coste, J., Michot, J. L., et al. (1999). Apolipoprotein E phenotypes in demented and cognitively impaired patients with and without cerebrovascular disease. *European Journal of Neurology*, 6(4), 415–421.
- von Campenhausen, S., Bornschein, B., Wick, R., BA¶tzel, K., Sampaio, C., Poewe, W., et al. (2005). Prevalence and incidence of Parkinson's disease in Europe. *European Neuropsychopharmacology*, 15(4), 473–490.
- Walker, Z., Allen, R. L., Shergill, S., Mullan, E., & Katona, C. L. (2000). Three years survival in patients with a clinical diagnosis of dementia with Lewy bodies. *International Journal of Geriatric Psychiatry*, 15(3), 267–273.
- Warrington, E. K. (1975). The selective impairment of semantic memory. *Quarterly Journal of Experimental Psychology*, 27(4), 635–657.
- Warrington, E. K. (1981). Concrete word dyslexia. *British Journal of Psychology*, 72(2), 175–196.
- Warrington, E. K., & Shallice, T. (1979). Semantic access dyslexia. *Brain*, 102, 42–63.
- Weisman, D., & McKeith, I. (2007). Dementia with Lewy bodies. *Seminars in Neurology*, 27(1), 42–47.
- Wetterling, T., Kanitz, R. D., & Borgis, K. J. (1996). Comparison of different diagnostic criteria for vascular dementia (ADDTC, DSM-IV, ICD-10, NINDS-AIREN). *Stroke*, 27(1), 30–36.
- Whitwell, J. L., Anderson, V. M., Scapill, R. I., Rossor, M. N., & Fox, N. C. (2004). Longitudinal patterns of regional change on volumetric MRI in frontotemporal lobar degeneration. *Dementia and Geriatric Cognitive Disorders*, 17(4), 307–310.
- Wilson, S. M., Brambati, S. M., Henry, R. G., Handwerker, D. A., Agosta, F., Miller, B. L., et al. (2009). The neural basis of surface dyslexia in semantic dementia. *Brain*, 132(Pt 1), 71–86.
- Woollams, A. M., Ralph, M. A., Plaut, D. C., & Patterson, K. (2007). SD-squared: On the association between semantic dementia and surface dyslexia. *Psychological Review*, 114(2), 316–339.
- World Health Organization. (1992). *International classification of diseases*. Switzerland: Geneva.
- Zaccai, J., McCracken, C., & Brayne, C. (2005). A systematic review of prevalence and incidence studies of dementia with Lewy bodies. *Age and Ageing*, 34(6), 561–566.