The Cambridge Handbook of Communication Disorders

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Dementia and communication

Jamie Reilly and Jinyi Hung

15.1 Introduction

Human longevity is rapidly increasing across much of the industrialized world, and this changing mosaic of ageing has created unprecedented challenges for healthcare systems. We are now in the midst of a public health crisis with respect to the management of Alzheimer's disease and associated forms of dementia. Moreover, all demographic indicators predict an exponential growth of dementia over the next three decades as a large proportion of the world's population approaches late middle age (Hebert *et al.* 2001, 2004; Alzheimer's Association 2010). In recognition of this looming epidemic, legislators have recently implemented a number of initiatives targeting dementia prevention and management. Much of this effort has focused on promoting advances in protein structure, genetics and molecular biomarkers. Recent advances in each of these domains hold great promise for identifying new drug targets and/or vaccines. Nonetheless, the state of cognitive rehabilitation for this rapidly growing segment of our society remains fundamentally limited. This is especially true with respect to disorders of speech, language and human communication.

It is now clear that we must find ways to promote functional independence and forgo institutionalized care (e.g. skilled nursing) for the many millions of adults who will soon be directly impacted by dementia. Communicative disorders are among the most functionally debilitating symptoms incurred in many different forms of dementia. Thus, the development of effective speech-language interventions is of paramount

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importance. It is paradoxical that formal training in dementia is not yet mandated within the graduate clinical curricula of many national speech-language therapy organizations (American Speech-Language-Hearing Association 2008). Moreover, many speech-language clinicians continue to espouse outmoded ideas about the potential for learning in the dementias (e.g. 'What's the use when they are getting worse anyway?'). A number of recent studies have questioned this deeply ingrained view by showing that certain techniques can indeed promote functional communication for patients with various forms of dementia, especially when paired with common drug adjuvants (Grandmaison and Simard 2003; Boyle 2004; Fridriksson *et al.* 2005; Gonzalez-Rothi *et al.* 2009).

We have learned a number of important lessons from studies of language learning in the dementias to date. Perhaps most importantly, treatment effectiveness is moderated by disease aetiology. Techniques that 'work' for promoting communication in one dementia variant (e.g. Alzheimer's disease) are likely to have very limited effectiveness for other dementia variants (e.g. frontal variant frontotemporal dementia). Therefore, variability across the dementia subtypes demands an aetiology-specific treatment approach. The dominant rate-limiting factor undermining treatment development is a lack of understanding about the unique cognitive profiles of the dementia subtypes and their relation to anatomical distributions of cortical atrophy. Here we address this issue by providing an overview of the cognitive communication profiles of several of the most common forms of dementia. These forms are Alzheimer's disease, frontotemporal dementia, Lewy body spectrum disease and vascular dementia. We issue a caveat that this review is necessarily highly selective with respect to both its breadth and depth. Space restrictions preclude coverage of all forms of dementia (e.g. HIV-AIDS dementia complex, Wernicke–Korsakoff dementia, dementia pugilistica) and all cognitive domains within the populations we do address. Thus, we focus specifically on speech, language and the essential cognitive processes that support human communication.

15.2 What is dementia?

Ageing refers to a constellation of physical, psychological and social changes in a person over time. Structurally, the ageing brain decreases in volume significantly across the lifespan (Drag and Bieliauskas 2010). However, these changes in structure are often compensated for by functional reorganization (Cabeza 2002). Healthy cognitive ageing is associated with preserved social and occupational functioning (Rowe and Kahn 1997), but for many people this trajectory is compromised. The transitional state between healthy normal ageing and frank dementia is known as mild cognitive impairment (MCI). Accordingly, a person with MCI is often subjectively classified as not normal but not demented.

MCI can persist in a chronic, relatively stable form for many years. However, a proportion of MCI cases show evidence for progressive deterioration. These patients experience a shift from MCI to dementia. Current diagnostic criteria for MCI include (1) objective or subjective concern regarding a change in cognition, (2) impairment in one or more cognitive domains, (3) preservation of independence in functional abilities and (4) no evidence of dementia. The incorporation of biomarkers is also suggested, especially in the diagnosis of MCI due to Alzheimer's disease (Albert *et al.* 2011). Clinical diagnostic criteria delineate three distinct subtypes of MCI (i.e. amnestic, multiple cognitive domains and single non-memory domain) based on the dominant presenting cognitive impairment. Amnestic MCI presents as a dominant impairment of episodic memory (Petersen *et al.* 2001). Postmortem histological studies have shown that this particular MCI subtype has the highest likelihood of evolving to Alzheimer's disease (Jicha *et al.* 2006). MCI can also manifest as a more heterogeneous condition involving the compromise of other cognitive functions such as attention, language and visuospatial functioning (Petersen 2004; Winblad *et al.* 2004; Petersen and Negash 2008). Language may appear to be relatively unaffected in the amnestic variant of MCI. However, subtle impairments are often detectable when the complexity of the linguistic demands is increased (Fleming and Harris 2008).

MCI will often gradually evolve to a frank form of dementia over time. From a probabilistic standpoint, MCI is most likely to evolve to Alzheimer's disease. However, probabilistic reasoning and a chronic lack of specificity has led to the common misconception that dementia and Alzheimer's disease (AD) are synonymous. AD is indeed the most common form of dementia, accounting for approximately 60 per cent of all new cases (Alzheimer's Association 2010). However, there exist a number of non-Alzheimer's dementia variants. These variants are classified by means of their dominant protein aggregations (e.g. Lewy body dementia involves synucleinopathy, while frontotemporal dementia involves tauopathy), or metabolic and systemic causes (e.g. vascular dementia, Wernicke–Korsakoff dementia, HIV-AIDS dementia complex). Each of these dementia subtypes manifests in a unique cognitive profile. However, there are also many overlapping phenotypic similarities that complicate differential diagnosis. Thus, 'dementia' is a non-specific umbrella term describing a set of broad features which the many disparate subtypes share in common.

Two dominant systems of criteria exist for establishing a diagnosis of dementia: the International Classification of Diseases (ICD-10; World Health Organization 1993) and the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV-TR; American Psychiatric Association 2000). According to the ICD-10, a diagnosis of dementia is appropriate in the context of two or more cognitive declines (e.g. memory, judgement, thinking, learning, orientation, language, comprehension or calculation)

that compromise one's daily functioning significantly. The DSM-IV-TR suggests dementia is a gradual and progressive memory disturbance with one or more of the following: aphasia, apraxia, agnosia and dysexecutive disorder without the occurrence of other reversible causes. As the reader may surmise, these criteria are necessarily broad. There is no single objective measure, either psychometric or physiological, that can definitively confirm the presence of dementia. Instead, diagnosis is a probabilistic process whereby clinicians must weigh evidence from a variety of sources, including behavioural testing, neuroimaging, family history and other biomarkers (e.g. cerebrospinal fluid proteins) (Dubois *et al.* 2007; Ewers *et al.* 2011; Holtzman *et al.* 2011).

Speech and language characteristics provide powerful diagnostic markers that can aid in detecting the presence of dementia (i.e. sensitivity). Consideration of unique speech and language impairments can also aid in the more challenging task of discriminating between dementia subtypes (i.e. specificity). We now turn to discussion of the profiles of communicative impairment associated with a range of dementia subtypes.

15.3 Alzheimer's disease

Our most common association of AD is that of a disorder of impaired episodic memory (i.e. recall of specific events). Episodic memory impairment is one of the dominant cognitive symptoms during the mild to moderate stages of AD prior to the onset of personality changes and a constellation of other perceptual and linguistic problems. AD has historically been classified as an amyloidopathy in that its phenotype has been linked to depositions of the protein beta-amyloid. Although recent work has implicated a range of other proteins in AD (e.g. tau), much of our understanding about the nosology and progression of the disease has been informed by studies that track the progression of amyloid plaque and tangle pathology.

AD tends to follow a canonical progression with respect to the distribution and sequence of brain regions commonly impacted (McKhann *et al.* 1984, 2011). This progression is reflected in the Braak staging system (Braak and Braak 1997; Braak *et al.* 2006). This system posits the presence of a series of discrete stages of AD characterized by a period of clinically silent degradation in structural connectivity between the medial temporal lobe and the cortex proper (i.e. transentorhinal stage), followed by degradation of the hippocampal formation and later by diffuse plaque and tangle pathology in many other cortical and subcortical regions (Holtzman *et al.* 2011).

The preclinical or prodromal stage of AD may extend for many years prior to the onset of frank dementia symptoms (Morris 2005; Sperling *et al.* 2011). One of the challenges for medical management of AD is to establish precise differential diagnosis and begin treatment (both pharmacological

and cognitive) prior to the onset of debilitating cognitive impairments (Parasuraman and Haxby 1993; Perry and Hodges 1999; Belleville *et al.* 2007). The most recent clinical criteria for AD diagnosis reflect a range of signs and symptoms, and further redefine the classification of the disease (i.e. probable AD dementia, possible AD dementia, and probable or possible AD dementia with evidence of the AD pathophysiological process). They also emphasize the incorporation of fluid or imaging biomarkers of the underlying disease state (McKhann *et al.* 2011).

Cognitive and linguistic impairments in AD are linked to a combination of diffuse synaptic loss and deposition of neuritic plaques within specific regions of the cortex (e.g. hippocampus, visual association cortex). In addition to gross structural grey and white matter loss, AD is also associated with a depletion of acetylcholine, a neurotransmitter that is essential for learning and memory encoding (Holtzman *et al.* 2011). Impairment in recent episodic memory, linked primarily to disconnection and atrophy of the medial temporal lobe, is one of the most common symptoms of AD. As hippocampal damage worsens, patients experience worsening anterograde amnesia, a condition characterized by a failure to effectively encode new memories (Nestor *et al.* 2006). During the early stages of AD patients tend to show a temporal gradient in forgetting that is characterized by worse recall for recent, relative to remote, episodic memories. For example, a patient might better recall details of her wedding day 50 years ago than the physician she met 15 minutes ago. Patients during later-stage AD typically show deficits in other forms of memory, including working memory and semantic memory. In addition to these associated amnestic impairments, AD also compromises a range of other cognitive domains related to language perception, executive function, attention, working memory and visuospatial abilities (Lambon Ralph *et al.* 2003; Nestor *et al.* 2004; Stopford *et al.* 2007). One useful diagnostic shortcut for detecting the presence of AD is that clinicians should look for the three As: Aphasia, Agnosia and Apraxia (but see Kramer and Duffy 1996).

Anomia (i.e. impaired naming) is a core feature of AD. Patients tend to show deficits in naming proper nouns and have also shown category-specific naming impairments for biological natural kinds relative to manufactured artefacts (Garrard *et al.* 1998; Capitani *et al.* 2003). In addition, patients tend to show a pattern of graceful degradation that is apparent in errors such as the overuse of superordinate category labels (e.g. animal, thing) in place of more descriptive basic or subordinate terms (e.g. dog, spaniel) (Martin and Fedio 1983; Hodges *et al.* 1992a).

The aetiology of the naming impairment in AD remains controversial. Some have attributed it to bottom-up degradation of a hierarchically organized semantic memory system (e.g. spaniels → dogs → mammals → animals → things). This claim receives converging support from impairments on other semantic tasks in AD, such as word-picture matching, priming and semantic fluency naming (Hodges *et al.* 1992a;

Rosser and Hodges 1994; Cerhan *et al.* 2002; Henry and Crawford 2004; Henry *et al.* 2004; Rogers and Friedman 2008). Another strong source of evidence in support of a semantic degradation account of anomia in AD is that patients tend to show strong correlations between 'naming and knowing' (Hodges *et al.* 1996). In a classic study, Hodges *et al.* (1996) demonstrated that the quality of concept definitions was worse for items patients could not name relative to successfully named targets. As counterpoint, others have argued that anomia in AD has a basis in impaired linguistic and/or perceptual access to semantic knowledge (Nebes *et al.* 1984, 1989). There exist complex and compelling arguments for both points of view. However, it is undeniable that deficits in visual perception (e.g. agnosia) and lexical retrieval moderate naming ability in AD and that these factors must be considered when planning treatments (Harnish *et al.* 2010).

As a general heuristic, output phonology, morphology and syntactic processing tend to remain intact relative to the massive loss of semantic memory and naming ability in AD. There are, however, noteworthy exceptions. Croot *et al.* (2000) reported non-fluent, agrammatic production in a series of AD patients with atypical perisylvian atrophy (see also Biassou *et al.* 1995). Barring exceptions such as these, people with AD tend to produce narrative discourse that is morphosyntactically well-formed but impoverished in terms of semantic content (for a review, see Almor *et al.* 1999). Patients tend to revert to over-learned phrases and idioms (e.g. 'You know, it's that thing') as ineffective means of circumlocution. Reductions in mean length of utterance (MLU) syntactic complexity and idea density are also common macro-scale features of discourse in AD (Almor *et al.* 1999). In summary, narrative discourse in AD is in many ways consistent with the lyric of the Talking Heads song *Psycho Killer*, 'You're talking a lot, but you're not saying anything' (Byrne *et al.* 1977).

Sentence comprehension is often compromised in AD, and there is much debate as to the cause(s) of this impairment. Causes include impairments in processing the meaning of single words and in comprehending the syntactic rules that govern word order. Patients with AD do show worse impairment as syntactic complexity increases. However, this relationship is non-linear and open to alternative explanations (Kempler *et al.* 1998). Sentence comprehension difficulties in AD may also be attributable to limitations in working memory. Although this issue remains controversial, the evidence to date seems to favour the hypothesis that sentence comprehension problems are related to impaired working memory rather than to a domain-specific syntactic impairment.

In addition to a range of frank impairments in single word, sentence and narrative comprehension, AD also compromises many higher-level linguistic processes linked to figurative language. Patients with AD commonly experience impairments in comprehension of non-literal language such as metaphor, idiomatic expressions, proverbs, irony, humour

and sarcasm (see Rapp and Wild (2011) for a review). These difficulties manifest as overly concrete and rigid interpretation of word meaning and consequent failure to grasp nuanced messages (see also Cummings 2007a). However, further investigation is still needed to expand current findings.

15.4 Frontotemporal dementia

Frontotemporal dementia (FTD) is a non-Alzheimer's dementia with an onset approximately a decade earlier than that of AD. The onset of FTD follows a roughly normal distribution with a mean early in the sixth decade of life and tapering incidence in older age. This Gaussian/normal distribution of onset distinguishes FTD from other forms of dementia such as AD that show a linear increase in risk as a function of advancing age (Ratnavalli *et al.* 2002). Neurodegeneration in FTD has been linked to proteins such as tau, ubiquitin and TDP-43 (Bian and Grossman 2007; Seelaar *et al.* 2008). Tau abnormalities, in particular, have been linked to neuronal microtubule collapse and subsequent cell death (but see Avila *et al.* 2002). Accordingly, FTD has been classified within a family of dementias known as tauopathies which also include motor neuron disease, cortical basal degeneration and progressive supranuclear palsy (Kertesz *et al.* 2000; McKhann *et al.* 2001; Boxer and Miller 2005).

FTD is a histological designation that subsumes a variety of behavioural subtypes (i.e. phenotypes). These behavioural presentations are linked to the primary distribution of atrophy incurred during the disease course. For unknown reasons, cortical atrophy remains relatively circumscribed within specific regions of the cortex during early stages of FTD. Patients commonly show hemispheric asymmetry in disease progression as well as unique patterns of lobar degeneration within each hemisphere. For example, one variant of FTD (i.e. semantic dementia) tends to produce atrophy most evident in the left lateral inferior temporal lobe (Snowden *et al.* 1989). We focus our discussion to follow on three of the most commonly recognized FTD syndromes: progressive non-fluent aphasia, semantic dementia and frontal variant FTD (Gorno-Tempini *et al.* 2011). A fourth syndrome, logopenic progressive aphasia, has also been identified. However, the most recent postmortem clinicopathological correlation studies have shown that this subtype may in fact more commonly represent an atypical form of AD (Mesulam *et al.* 2008).

15.4.1 Progressive non-fluent aphasia

Progressive non-fluent aphasia (PNFA) profoundly compromises a person's ability to produce fluent and grammatically well-formed speech (Gorno-Tempini *et al.* 2004). The most recent core diagnosis of PNFA includes

agrammatism and effortful speech. Patients with PNFA invariably show slowed speech output and articulatory struggles along with the presence of restarts, repeated syllables and phonemic paraphasias (Ash *et al.* 2004, 2010). Additional supportive features include difficulty comprehending syntactically complex sentences, spared single word comprehension and spared object knowledge (Gorno-Tempini *et al.* 2011). PNFA often evolves to complete mutism (Gorno-Tempini *et al.* 2004; Gunawardena *et al.* 2010; Rabinovici and Miller 2010).

The aetiology of the output impairment associated with PNFA remains controversial. Some have noted the presence of apraxia of speech in PNFA, which compromises motor aspects of production (Gorno-Tempini *et al.* 2004; Ogar *et al.* 2007). Others have argued that many of the deficits in production in PNFA share higher-level linguistic bases such as deficient phonological encoding, agrammatism and gross executive function limitations (Libon *et al.* 2007; Knibb *et al.* 2009; Ash *et al.* 2010; Gunawardena *et al.* 2010). Supporting this latter view, there is a tendency for patients with PNFA to produce reduced frequency of grammatically complex sentences (Ash *et al.* 2010; Gunawardena *et al.* 2010).

In our own work we have found that patients with PNFA tend to experience severe anomia, and these effects may be worse for manufactured artefacts than for animals or other biologically natural kinds (Reilly *et al.* 2011a). Others have reported that the naming impairment in PNFA is worse for verbs relative to nouns (Hillis *et al.* 2004b). In addition to category effects in naming, we also found evidence of verbal and non-verbal semantic impairment in PNFA, suggesting that difficulties incurred by these patients transcend modality-specific impairments of language (but see Kempler and Goral 2008).

Patients with PNFA typically have difficulties in comprehending syntactically complex sentences in the context of relatively intact single word recognition (Rabinovici and Miller 2010). A definitive basis for this impairment is elusive. Some have argued the comprehension impairment is indeed related to a grammatical/syntactic deficit (Ash *et al.* 2010; Gunawardena *et al.* 2010). Others have argued that sentence comprehension deficits are more likely attributable to impaired working memory (Grossman and Moore 2005; Peelle *et al.* 2008).

15.4.2 Semantic dementia

Semantic dementia (SD) is a variant of FTD associated with progressive bilateral degeneration of the temporal lobes (Hodges 2001; Grossman *et al.* 2002; Rabinovici and Miller 2010). During the early stages of SD, cortical atrophy is often asymmetric (left hemisphere damage is greater than right) and most prominent in anterolateral aspects of the temporal lobe (Peelle and Grossman 2008; Rabinovici and Miller 2010). SD is distinct from AD in that the pathology typically spares medial temporal

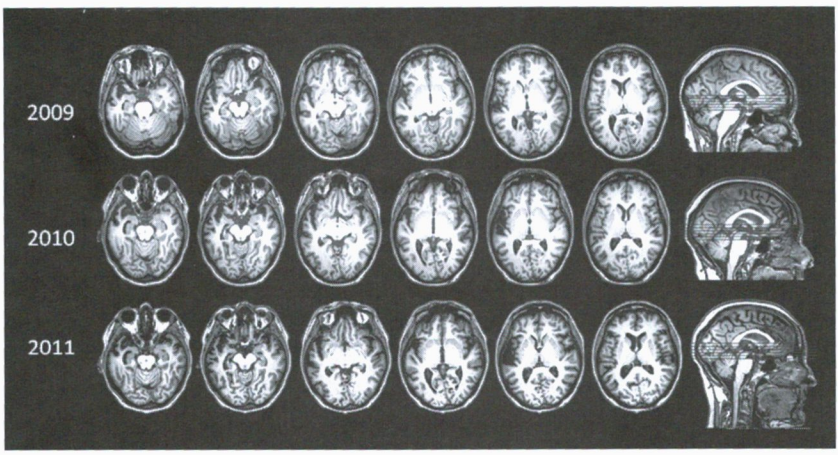

Figure 15.1 Successive MRI scans of a patient with semantic dementia.

structures that support episodic memory encoding and new anterograde learning. Figure 15.1 represents the distribution of atrophy in a patient with SD scanned successively over a 3-year period.

The most pronounced feature of SD is its associated degradation of semantic knowledge. Patients often display a homogeneous loss of semantic knowledge that transcends representational modality (e.g. written or spoken text, pictures, environmental sounds) and mode of input or output (e.g. comprehension versus expression) (Bozeat *et al.* 2000). During the progression of the disease, patients with SD show relative preservation of anterograde memory for recent day-to-day events as well as preserved sensory processing (these traits are commonly disturbed in AD). It is not uncommon for patients to successfully discriminate pictures or sounds (same/different) or complete other perceptual matching tasks with relative ease. Output phonology, syntax and speech articulation are also relatively preserved during much of the course of SD (Jefferies *et al.* 2006). Preserved single word repetition and fluent speech are core features of the disease (Neary *et al.* 1998). This range of apparently preserved abilities is in stark contrast to the profound loss of conceptual knowledge that underlies word and object meaning.

Warrington (1975) famously described the selective impairment of semantic memory we now regard as semantic dementia (Snowden *et al.* 1989). In the decades following Warrington's seminal article, SD has provided a powerful, yet highly controversial, lesion model for parsing the organization of human conceptual knowledge. SD has also informed cognitive science about the interplay between disturbed semantic knowledge and its effect on other cognitive processes (e.g. word recognition, colour perception) (Reilly *et al.* 2007a, 2007b, 2011b; Reilly and Peelle 2008).

Patients with SD usually produce fluent but empty speech with frequent semantic paraphasias (Grossman and Ash 2004; Kempler and Goral 2008; Kertesz *et al.* 2010). Spontaneous speech is often characterized by profound anomia with better performance on typical, familiar and high-frequency words (Patterson 2007; Meteyard and Patterson 2009). Patients also commonly revert to deictic phrases, idioms and non-specific names (e.g. things, stuff) (Grossman and Ash 2004). Importantly, there appears to be a strong correlation in SD between 'naming and knowing' with patients unable to produce words for which their conceptual knowledge is disrupted. Patients tend to show minimal benefits from overt semantic cueing and demonstrate minimal priming effects (Reilly *et al.* 2005). The naming impairment in SD is one manifestation of a more pervasive loss of object knowledge that is also evident in non-verbal domains (e.g. demonstrating object function and use, categorizing pictures of objects) (Bozeat *et al.* 2000; Adlam *et al.* 2006).

SD is associated with severe impairments in single word comprehension, paralleling the impairment in naming (Reilly *et al.* 2007a; Gorno-Tempini *et al.* 2011). Comprehension deficits are moderated by factors such as disease severity, concept familiarity, word frequency and item typicality (Adlam *et al.* 2006). Sentence processing impairments are also common in SD, and these deficits are most often attributed to a semantic locus relative to other cognitive processes (e.g. agrammatism or working memory deficiencies) (Gorno-Tempini *et al.* 2004; Peelle *et al.* 2008).

One of the most ubiquitous and striking symptoms of SD is its associated pattern of reading impairment, known as surface dyslexia (Woollams *et al.* 2007). Surface dyslexia is remarkable for the preserved ability to read aloud words with transparent orthography in the context of impaired reading of orthographically irregular words (e.g. sew, yacht). A similar error pattern of orthographic regularization known as surface dysgraphia is also evident in written production (Grossman and Ash 2004; Wilson *et al.* 2009; Kertesz *et al.* 2010).

Dual route models of reading provide a compelling account of surface dyslexia in SD. That is, patients revert to preserved phonological knowledge to rigidly convert graphemes to phonemes. Healthy adults supplement this phonological process using word meaning and whole-word recognition. This putative semantic route is, however, unavailable in the context of SD (Woollams *et al.* 2007). Further evidence for this pattern was derived from Japanese patients with SD who were asked to read aloud words in the two orthographic systems of Japanese, i.e. Kana and Kanji. Kana is transparent in the relationship between orthography and phonology whereas the pronunciation of Kanji characters is context driven. Japanese patients with SD consistently performed well when reading Kana. However, they were selectively impaired in reading Kanji with atypical correspondences (Fushimi *et al.* 2009).

15.4.3 Frontal variant FTD/behavioural variant FTD

The frontal/behavioural variant of frontotemporal dementia (fvFTD) is not typically associated with the profound communicative impairments that are evident in SD and PNFA. The anterior cingulate, insular and orbitofrontal cortex are most affected in fvFTD (Rosen *et al.* 2002; Seeley *et al.* 2008). fvFTD is also sometimes referred to as social dysexecutive disorder due to its range of progressive deficits in executive functioning, inhibitory control and emotional regulation. A recent proposal by the International Behavioral Variant FTD Criteria Consortium (Rascovsky *et al.* 2011) introduces a hierarchy of diagnostic certainty including possible, probable and definite fvFTD. The diagnosis of possible fvFTD requires three random presentations of six behavioural/cognitive symptoms: disinhibition, apathy/inertia, loss of sympathy/empathy, perseverative/compulsive behaviour hyperorality/dietary changes and dysexecutive neuropsychological profile. The diagnosis of probable fvFTD further includes evidence of daily functional decline and pathological support from imaging results. fvFTD with definite pathology is only to be applied to those patients who show clinical syndromes with clear histopathological evidence or a known pathogenic mutation.

Unlike previous consensus criteria, this revised proposal emphasizes the distinctive behaviours of early stages of fvFTD and attempts to increase the sensitivity of diagnosis. Its reliability and specificity await further investigation (Rascovsky *et al.* 2007, 2011). Previous diagnostic criteria also list speech and language as supportive features of fvFTD including altered speech output, stereotypy of speech, echolalia, perseveration and mutism (Neary *et al.* 1998). These impairments are often apparent at the level of discourse and connected speech (Libon *et al.* 2007).

Patients with fvFTD are commonly impaired in measures of semantic and phonemic verbal fluency (e.g. 'Tell me as many animals as you can in one minute') (Libon *et al.* 2009). Many of these limitations have been attributed to deficits in switching and task vigilance that are classically subsumed under the domain of executive functions (Libon *et al.* 2009). Patients with fvFTD do not commonly experience the severity of anomia that is present at the single word level in PNFA or SD. However, deficits in expressive language are indeed present at the discourse level. Ash *et al.* (2006) examined aspects of discourse such as narrative coherence and maintenance of theme as patients with fvFTD narrated the story depicted in the wordless children's book *Frog, where are you?* (Mayer 1969). These authors found that fvFTD patients were able to find words to describe a picture, but that their narratives lacked connections to bind information into a coherent story, reflecting difficulty organizing their narratives (Ash *et al.* 2006).

Patients with fvFTD display relatively preserved comprehension of single words but show deficits for tasks that load on executive and working memory demands. This includes manipulations of syntactic and narrative

complexity (Peelle and Grossman 2008). Patients have also been reported to have difficulties in understanding non-literal language critical for the appreciation of humour, irony and metaphor (Kosmidis *et al.* 2008; Kipps *et al.* 2009). As a result, fvFTD language comprehension often assumes a degree of rigid literality.

15.5 Vascular dementia

Vascular dementia (VaD) is a common form of dementia caused by a variety of cerebrovascular pathologies, including multiple strokes and small vessel ischaemic disease. The phenotype of VaD is moderated by a number of factors including nature of the arterial disease (i.e. small or large), type of stroke (i.e. ischaemic or haemorrhagic), site of lesion (e.g. cortical or subcortical), number of infarcts and other comorbid health factors (Román *et al.* 1993; Jellinger 2008). There exist no less than four distinct sets of diagnostic criteria for VaD in active use today. These include 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 (Román *et al.* 1993), the Diagnostic and Statistical Manual of Mental Disorders (American Psychiatric Association 1994) and the International Classification of Diseases (World Health Organization 1993). The variability of these diagnostic criteria reflects the heterogeneous nature of the disease.

Diagnostic specificity for VaD is complicated by high comorbidity with other neurological disorders such as Alzheimer's disease and traumatic brain injury (O'Brien *et al.* 2003; Nagata *et al.* 2007; Benisty *et al.* 2008). A number of studies have recently contrasted language and amnestic impairment in VaD and AD (Lafosse *et al.* 1997; Almkvist *et al.* 1999; Graham *et al.* 2004). Although some of these studies reveal differences between VaD and AD, there is not universal agreement as to the discriminative neuropsychological features of these disorders. The differential diagnosis of VaD is further complicated by the fact that there is no definitive threshold for dementia in the presence of multiple strokes. These challenges in diagnostic specificity are illustrated by a simple hypothetical example of two patients with chronic vascular disease. Patient A has sustained multiple ischaemic injuries to the left inferior frontal cortex resulting in non-fluent aphasia. Patient B has sustained diffuse white matter damage resulting from small vessel ischaemic disease. Patients A and B both satisfy many criteria for VaD, and yet they are likely to present with two very different behavioural profiles.

Despite inherent variability introduced by the diffuse nature of the human cerebral vasculature, there are some common features of VaD. Vascular damage tends to prominently affect the white matter, resulting

in a condition known as leukoaraiosis (van Gijn 1998). White matter damage (as incurred in HIV-AIDS dementia) is associated with a specific range of cognitive deficits including slowed information processing, impaired working memory, poor sequencing, lack of inhibitory control, and a number of related impairments in executive functioning and attention (Starkstein *et al.* 1996; Mendez *et al.* 1997; Yuspeh *et al.* 2002; McGuinness *et al.* 2010). These deficits are commonly seen in VaD and many have been hypothesized to provide a substrate for associated impairments in language comprehension and expression.

One common finding regarding expressive language in VaD is that patients tend to show reduced verbal fluency relative to other dementia control groups (Starkstein *et al.* 1996; Lafosse *et al.* 1997). It is suggested that executive dysfunction, especially mental processing speed, contributes to decrements in fluency in VaD (Lafosse *et al.* 1997; Jones *et al.* 2006). For non-aphasic patients with VaD, naming is not typically as impaired as that of AD (Lukatela *et al.* 1998; Graham *et al.* 2004). Very few studies have investigated the integrity of receptive language in VaD (Desmond *et al.* 1999; Vuorinen *et al.* 2000; Desmond 2004). While comprehension impairments have indeed been reported, the basis of these deficits remains unclear and potentially multifactorial.

15.6 Synucleinopathy spectrum disorders

Parkinson's disease dementia (PDD) and Lewy body dementia (LBD) represent a spectrum of dementias known as synucleinopathies. In both conditions, large aggregations of destructive alpha-synucleinated proteins known as Lewy bodies accumulate in specific brain regions. When Lewy bodies destroy more than about 80 per cent of the dopaminergic cells within the substantia nigra, patients tend to show the overt motor symptoms of classical Parkinson's disease (Louis and Frucht 2007). In contrast, Lewy body deposition in the cortex produces a different syndrome known as LBD. In reality, these spectrum disorders are not always easily disentangled due to the fact that Lewy body damage tends to affect both the substantia nigra and the cortex in both conditions. For this reason, neurologists commonly employ what is known as the 'one year rule' when establishing a differential diagnosis of PDD or LBD. A diagnosis of PDD is appropriate when cognitive symptoms emerge within the context of a movement disorder lasting more than one year. By contrast, a diagnosis of LBD applies when movement disorders emerge in the context of a pre-existing dementia. When we describe the nature of language and communication in PDD and LBD below, we operate under an assumption that these syndromes have similar histopathological courses and many shared traits (McKeith 2000). It should be noted, however, that the lumping of PDD and LBD as spectrum disorders is not a universally accepted

practice (Revuelta and Lippa 2009). We will describe these clinical populations as unique entities while also acknowledging a similarity bias.

15.6.1 Parkinson's disease dementia

Parkinson's disease (PD) is a chronic neurodegenerative condition that has historically been regarded as a movement disorder. Patients with PD commonly present with cardinal motor symptoms such as bradykinesia, tremor, rigidity and postural instability (Román *et al.* 2004; Bartels and Leenders 2009). However, a growing body of research implicates a range of cognitive impairments that are associated with non-demented PD, suggesting that PD cannot be classified exclusively as a movement disorder (Lewis *et al.* 2005; Williams-Gray *et al.* 2007; Mamikonyan *et al.* 2009; Aarsland *et al.* 2010; Kehagia *et al.* 2010). For example, one pooled analysis showed that 26 per cent of 1,346 patients with PD satisfied diagnostic criteria for mild cognitive impairment (MCI) due to impairments in a variety of domains, including delayed recall, attention/executive functioning, language and visuospatial functioning (Aarsland *et al.* 2010). The incidence of MCI in the larger population of patients with PD remains unclear.

Another factor that remains unclear is the rate/risk of evolution to dementia in PD. Some studies have reported that up to 80 per cent of patients satisfy criteria for dementia one decade after motor symptom onset. Others have reported more conservative estimates of 48 per cent (Emre *et al.* 2007). In either case, when dementia emerges within the context of a patient with non-demented PD, that patient may be classified as having PDD. In addition to a prior diagnosis of PD, diagnostic criteria for PDD require the gradual decline in more than one cognitive domain (i.e. attention, executive functions, visuospatial functions, memory or language) and the presence of at least one behavioural symptom (i.e. apathy, personality/mood changes, hallucinations, delusions or excessive daytime sleepiness) (Emre *et al.* 2007).

The communicative profiles of AD and FTD have been investigated extensively to date. However, the same cannot be said of PDD. A few studies have contrasted PDD with AD. The most common finding is that patients with PDD tend to fare slightly better in terms of the severity of their expressive language impairments (Emre 2003; Emre *et al.* 2007). Most studies link impairments in expressive language in PDD to processing limitations, some of which are compounded by concurrent motor impairments (e.g. working memory compounded by slowed speech output). Impaired verbal fluency is common in PDD and is often worse than that of AD (Henry and Crawford 2004). PDD is also associated with naming impairment, with some studies reporting worse performance for verbs and actions relative to nouns and objects (Cotelli *et al.* 2007; Murray 2008; Rodriguez-Ferreiro *et al.* 2009).

PDD presents a unique set of challenges in terms of comprehension impairment. Sentence comprehension deficits are common in PDD, especially for utterances with non-canonical syntactic structures (Grossman *et al.* 1991; Goetz *et al.* 2008). The nature of this impairment is controversial and follows similar lines to that seen in aphasia and also AD (i.e. 'Is the problem grammatical or attributable to a more general processing impairment?'). Some have argued for the presence of a grammatical impairment based on selective deficits in understanding specific syntactic structures (Lieberman *et al.* 1992). However, the bulk of recent behavioural and neuroimaging evidence appears to favour a processing account that may affect syntax along with a variety of other supportive cognitive functions (Grossman *et al.* 2002). There has been relatively little work examining the integrity of semantic memory in PDD, and much remains unclear about the effects of extensive subcortical damage on semantic processing (Crosson 1992). In late stage PDD, patients may experience visual hallucinations and fluctuating periods of consciousness that compromise comprehension (Ibarretxe-Bilbao *et al.* 2010).

It is becoming increasingly apparent that non-demented PD compromises cognition and communication. Hypokinetic dysarthria is an associated motor speech disorder that tends to compromise speech intelligibility in PD, and micrographia (i.e. illegibly tiny writing) is also commonly seen in PD (Jankovic 2008). Patients with PD do not typically manifest the profound deficits in naming and language comprehension that characterize FTD or AD. Non-demented PD patients do, however, experience high-level language impairments, including comprehension and production of complex narrative. Many have argued that the primary basis for high-level language impairment in PD lies at the level of processing limitations. Executive dysfunction and working memory impairments are prominent in PD (with or without dementia), resulting in deficits in planning, inhibition, set-switching, goal-directed behaviour, strategy formation and working memory (Henry and Crawford 2004; Emre *et al.* 2007; Murray 2008).

Patients with PD commonly show diminished pitch and amplitude contours in their vocal output consistent with dysprosodia. These prosodic deficits are also accompanied by diminished variability of facial expressions, a phenomenon known as masked facies (Pell 1996; Pell and Leonard 2003; Pell *et al.* 2006). These characteristics often lead to the impression that a communicative partner with PD has undergone emotional blunting and loss of empathy. Indeed, PD is thought to compromise aspects of emotional communication both receptively and expressively (Pell and Leonard 2005).

Many of the deficits that are apparent in prosodic speech output in PD manifest as analogous impairments in comprehension. Patients with PD often experience insensitivity to two dissociable forms of prosody (i.e. affective and linguistic prosody) (Heilman *et al.* 1984; Hillier *et al.* 2007).

Deficits in affective prosody compromise a patient's ability to detect emotional content conveyed by fluctuations in pitch and amplitude of a speaker's voice. Such cues are essential for conveying many non-literal aspects of language and communication such as irony and humour, domains that can become difficult for patients with PD to appreciate. PD also compromises perception of linguistic prosody that is critical for disambiguating word meaning or grammatical class based on unique stress patterns. For example, 'content' can mean two different things depending on its syllabic stress. In the absence of additional contextual linguistic detail, PD patients often experience difficulties detecting such subtle acoustic cues (Kotz *et al.* 2009).

15.6.2 Lewy body dementia

Although prevalence estimates vary, it is believed that Lewy body dementia (LBD) is often under-diagnosed and may in fact represent the second most common dementia behind AD (Zaccai *et al.* 2005). Core clinical features of LBD include fluctuating attention, repeated visual hallucinations and spontaneous parkinsonism (McKeith *et al.* 1996). Suggestive features include rapid eye movement (REM) sleep behaviour disorder, profound neuroleptic sensitivity and low dopamine transporter uptake in the basal ganglia on functional neuroimaging. Patients with LBD commonly experience frequent falls and syncope, transient loss of consciousness, severe autonomic dysfunction, multimodal hallucinations, delusions, depression and paranoia (McKeith 2006; Weisman and McKeith 2007).

Cognitive dysfunction emerges early during the course of LBD. Executive dysfunction, inattention and visuospatial/visuoperceptual dysfunction are the most consistently reported complaints (Ferman and Boeve 2007). Working memory, episodic memory and semantic memory are also compromised in LBD, but the underlying mechanism and the impaired aspect of memory processing (e.g. encoding, retrieval or consolidation) remains unclear (Metzler-Baddeley 2007). The phenotype of LBD is often conceptualized as a multifactorial blend of visuospatial, amnestic and dysexecutive impairments (Doubleday *et al.* 2002). Although LBD is prominently associated with visual disturbance, primary language impairment is a less common presentation. Prominent language impairments include confabulatory speech production, incoherent conversation, irrelevant responses, anomia and reduced verbal fluency (Lambon Ralph *et al.* 2001; McShane *et al.* 2001; Doubleday *et al.* 2002; Ash *et al.* 2012).

The receptive language impairment associated with LBD has been linked to decrements in verbal working memory and executive functioning. That is, impairments often emerge beyond the single word level within the domain of narrative discourse. Patients with LBD experience difficulties in online processing of syntactically ambiguous sentences (Grossman

et al. 2012) and complex sentence structures (e.g. strategically padded sentences with additional prepositional phrases) (Gross *et al.* 2012).

15.7 Dementia and primary progressive aphasia

In a groundbreaking and highly cited series of works, neurologist Marcel Mesulam outlined formal criteria for the condition known as primary progressive aphasia (PPA) (Mesulam 1982, 2001, 2003, 2007). PPA manifests as a relatively focal impairment in the production and/or comprehension of language in the absence of frank dementia symptoms. PPA was accordingly described as 'slowly progressive aphasia without generalized dementia' (Mesulam 1982: 592). The most recent clinical diagnostic criteria for PPA delineate three distinct subtypes: non-fluent/agrammatic, logopenic and semantic PPA (Gorno-Tempini *et al.* 2011).

Unlike stroke aphasia, which tends to present as either stable or improving, PPA is by nature progressive. As speech and language impairments worsen across time, patients also tend to experience more of the classical symptoms of dementia, a stage recently termed PPA+ (Mesulam *et al.* 2003, 2009). Importantly, the progression from isolated speech-language impairment in PPA to more generalized dementia in PPA+ is continuous rather than punctuated. That is, no definitive threshold exists for when a person with PPA has crossed into the realm of dementia. For this reason, PPA has engendered great controversy (Snowden *et al.* 1989; Adlam *et al.* 2006).

PPA is the physical manifestation (i.e. phenotype) of one or more neurodegenerative processes. For example, one form of PPA (i.e. logopenic progressive aphasia) has been linked to primary AD pathology (Mesulam *et al.* 2008). PPA has been reported in autopsy-confirmed cases of FTD, LBD and VaD. However, the majority of PPA cases have been linked to the family of tauopathies that includes FTD (Grossman 2010). As such, some have argued that fluent variants of PPA do in fact represent early stage FTD.

Our discussion of PPA and dementia underscores a larger theoretical debate about the nature of aphasia in dementia. For over a century, aphasiology and neurology have been united in the belief that dementia does not fit easily within a classical cortical aphasia taxonomy (Wernicke 1874). One particularly influential dichotomy that has guided both assessment and treatment of aphasia in dementia is the distinction between disorders of access versus storage (Warrington and McCarthy 1983). Many speech-language pathologists, for example, operate under the assumption that dementia represents a core deficit of degraded storage whereas deficits in stroke aphasia are better characterized by impaired access to knowledge. This storage–access heuristic offers an intuitive and utilitarian framework for conceptualizing aphasia in dementia. However, this distinction is not without controversy, especially as it pertains to guiding one's treatment rationale (Rapp and Caramazza 1993; Reilly *et al.* 2011b).

15.8 Concluding remarks

In this chapter, we have presented an overview of communication in the dementias. The focus of the chapter has been on speech and language functioning in a small subset of dementia variants. Of course, this is problematic because human communication involves much more than speech and language. A comprehensive understanding of communicative impairment must also consider non-verbal expression and the ways that communication partners adapt to dementia. These aspects of communication in the dementias are still largely empirically uncharted. It is becoming increasingly evident that these gaps must be filled if we are to develop viable communicative interventions for the many millions of people living with dementia.