

# Neuropsychological Assessment of Primary Progressive Aphasia (PPA)

*Brianne M. Bettcher*

Memory and Aging Center, University of California San Francisco  
San Francisco, CA

*Virginia E. Sturm*

Memory and Aging Center, University of California San Francisco  
San Francisco, CA

*Financial Disclosure:* Brianne M. Bettcher is an Assistant Professor at the University of California San Francisco. Virginia E. Sturm is an Assistant Professor at the University of California San Francisco.

*Nonfinancial Disclosure:* Brianne M. Bettcher has previously published in the subject area. Virginia E. Sturm has previously published in the subject area.

## **Abstract**

*The goal of this article is to outline the utility of both language and non-language testing in making a diagnosis of logopenic, nonfluent/agrammatic, and semantic variant primary progressive aphasia PPA as well as delineate important behavioral and speech features that can be detected via clinical observation. We review speech/language presentations, non-language cognitive domains, and behavioral manifestations associated with each disorder. Patients with logopenic variant PPA evidence non-language cognitive impairments that include acalculia, phonological working memory deficits, and mild/variable difficulties with memory and visuospatial functions. In contrast, patients with nonfluent/agrammatic variant PPA display non-language impairments in executive functions, and show relative preservation of memory and visuospatial functions. Finally, semantic variant patients display behavioral changes in social comportment as well as non-language difficulties with category fluency and arithmetic facts; they display relative preservation, if not enhancement, of visuospatial functions. In summary, broad neural networks that support both language and non-language functions are affected in PPA syndromes, thus a comprehensive assessment of additional neuropsychological domains may aid in solidifying and subtyping PPA diagnoses.*

Primary progressive aphasia (PPA) are a group of neurodegenerative clinical syndromes, characterized by progressive, early deficits in language and/or speech functioning. Three syndromes are included in this umbrella category and include the logopenic, nonfluent/agrammatic, and semantic variants. Each syndrome is characterized by a prototypical phenotype that is driven by disruption in specific neural networks. As such, the presentation of each disorder includes an array of language, speech, cognitive, and behavioral disturbances that collectively contribute to and ultimately define the clinical picture seen in an evaluation. Whereas language deficits are the cardinal features of PPA syndromes, impairment in other cognitive and psychosocial domains is frequently evident, particularly with disease progression, rendering it important to conduct a full neuropsychological evaluation of patients whose symptoms suggest this diagnosis.

The goal of this article is to provide an overarching view of PPA clinical syndromes using the lens of neuropsychological assessment. Our perspective is based on recent research in the field and also emphasizes the neuropsychological assessment practices employed by our institution at the University of California, San Francisco Memory and Aging Center (UCSF MAC). We hope to convey the importance of (a) conducting comprehensive cognitive evaluations that assess multiple domains, both in terms of language and non-language measures; (b) characterizing relative strengths and weaknesses, through examination of performance; and (c) appreciating the neuroanatomy that

underlies each syndrome. Understanding syndrome-specific neuroanatomy is critically important as this knowledge may help to clarify the cognitive domains an evaluator can expect to be impaired and to delineate the domains that should remain relatively preserved or even enhanced until later stages of the disease course (Rogalski et al., 2011). The following sections will outline the utility of both language and non-language testing in making a diagnosis of logopenic, nonfluent/agrammatic, and semantic variant PPA as well as delineate salient behavioral and speech features that can be detected via clinical observation.

## **Logopenic Variant**

Logopenic variant PPA is a clinical syndrome typified clinically by frequent word-finding pauses, slow rate of speech, poor repetition of phrases, and syntactically simple conversational speech (Gorno-Tempini et al., 2008; Henry & Gorno-Tempini, 2010; Mesulam, Wieneke, Thompson, Rogalski, & Weintraub, 2012; Wilson et al., 2010). Although logopenic patients exhibit symptoms that may overlap with semantic variant, nonfluent/agrammatic variant, or even Alzheimer's disease, they also exhibit a striking constellation of cognitive and behavioral changes that can help to differentiate them from other syndromes. Their neuropsychological phenotype of logopenic PPA includes a wide range of impairments, including impoverished phonological working memory, phonological alexia, poor confrontation naming, and acalculia, with varying degrees of verbal memory impairments. This pattern of symptoms stems from a posterior brain etiology, driven by grey and white matter damage to the left temporoparietal junction, extending from the left posterior middle and superior temporal gyri to the inferior parietal lobule.

### **Clinical Observations**

Prior studies have identified several behavioral characteristics that may be observed in patients with logopenic variant, including apathy, anxiety, and mild irritability (Rohrer & Warren, 2010). Patients are often aware of their symptoms, and may report feeling embarrassed or concerned about their language difficulties in everyday life.

### **Speech-Language Functions in Logopenic Variant PPA**

Patients with logopenic variant PPA are often described as having an intermediate level of fluency relative to semantic variant (fluent) and nonfluent/agrammatic variant (nonfluent); their word finding difficulties will often be palpable in conversational speech, and are characteristically manifested by word finding pauses and circumlocutions in conversational speech, reductions in the number of words spoken per minute, and grammatically simple (but accurate) sentences.

When conducting an initial language assessment as part of a larger neuropsychological evaluation, identifying the presence *and* absence of specific speech and language symptoms are integral to solidifying a logopenic diagnosis, and are well-documented in the current PPA diagnostic criteria (Gorno-Tempini, 2011). Patients with logopenic variant display impaired single-word retrieval both on confrontation naming tasks as well as in speech samples (e.g., picture descriptions), and impaired repetition of sentences and phrases. In addition, phonologic errors in speech and on naming tasks are frequently observed. Although their paraphasias are typically characterized by phonological insertions or deletions that could be transcribed by the evaluator, intermittent or mild sound distortions may also be present. In contrast, these patients typically show preserved single-word comprehension and object knowledge, intact motor speech, and preserved grammatical structure in speech and writing.

### **Non-Language Cognitive Functions in Logopenic Variant PPA**

Although the earliest and most prominent cognitive symptoms reported by logopenic patients are with language functions, they evidence a stereotypical pattern of neuropsychological performance that extends beyond language deficits. Taking into consideration the anatomy and pathology associated with logopenic variant, one approach to clinical assessment is to consider "neighborhood signs," (i.e., clinical indicators that are also associated with the affected regions), including posterior temporal and inferior parietal (i.e., inferior parietal lobule; angular gyrus)

regions. The intraparietal sulcus and angular gyrus support processing of numerosity and complex calculations, and impairments in this cognitive domain are often reported fairly early in the clinical course. Careful evaluation of calculation ability, including the assessment of items requiring multiplication or complex addition is quite fruitful, and further differentiates logopenic variant from the other PPA syndromes. Furthermore, patients with logopenic variant may also display mild deficits on visuospatial tasks, including visual localization and construction (e.g., copying a complex figure).

One of the cardinal features of logopenic variant is phonological working memory difficulty, typically localized to inferior parietal regions and overlapping with the “phonological loop” construct referenced in Baddeley’s working memory models (Baddeley, 2003). As such, logopenic variant patients may perform poorly on measures that require maintenance of a brief verbal memory trace, such as Digit Span (forward digit span).

As the disease progresses to posterior temporal regions, connections to the posterior hippocampus are deleteriously affected. Congruent with this finding, patients with logopenic variant may display impoverished memory profiles on verbal list-learning tasks, with auditory memory typically worse than visual memory.

In terms of executive functions, whereas tasks mediated by dorsolateral prefrontal cortices are relatively spared in patients with logopenic variant, executive functions are driven by an extensive frontoparietal network, which may be impacted in these patients. It is not uncommon for patients to evidence mild impairments on measures of working memory and cognitive switching; however, severe impairments in executive functions early in the disease course are unusual. If a patient suspected of having logopenic variant displays striking executive dysfunction comparable to their language impairment on neuropsychological tests, this should raise diagnostic concern for an evaluator and may alter the differential diagnosis.

## ***Nonfluent/Agrammatic Variant***

Nonfluent/agrammatic variant primary progressive aphasia is a clinical syndrome characterized by progressive difficulty with syntactic aspects of language and/or motor speech impairment (Grossman et al., 1996; Grossman, 2012; Mesulam, 1982). These patients typically have slow, effortful, halting speech that may be agrammatic. Their neuropsychological profile typically includes more circumscribed impairments, with disproportionate difficulty on measures of executive functions and relative preservation of memory and visuospatial skills. These symptoms are consistent with the underlying anatomy of nonfluent/agrammatic variant, as left-lateralized atrophy in inferior frontal gyrus and posterior insula are pathognomonic of the syndrome, whereas posterior brain regions remain intact until late stages of the disease process.

### **Clinical Observations**

Patients with nonfluent/agrammatic variant typically display intact social skills, with social graces and interpersonal functioning often preserved in early stages of the disease course. Patients may be keenly aware of their performance on cognitive tasks and may express sadness and frustration about their speech impairment. Consistent with this observation, marked behavioral changes are atypical and uncommon manifestations of the syndrome, but mild disinhibition and apathy may appear as the disease progresses (Banks & Weintraub, 2008).

### **Speech-Language Functions in Nonfluent/Agrammatic Variant PPA**

The clinical phenotype of nonfluent/agrammatic variant is notable for agrammatism and/or speech production deficits, although not all patients show profound apraxia of speech early in the disease course. When patients do display apraxia of speech, sound distortions may range from subtle to prominent. Conversational speech may be further notable for altered prosody, resulting in phrases or sentences that lack changes in pitch or emotive emphasis.

In the context of formal language assessment, patients may have a slow rate of speech and make variable sound errors including distortions, substitutions, deletions, insertions, and transpositions of speech sounds on timed verbal agility tasks (e.g., tasks that require patients to repeat a single word as many times as they can in five seconds). Agrammatism may be apparent in speech or writing, and patients may use short, simple phrases that omit functor words (e.g., “the” and “to”). On tests of confrontation naming, word retrieval may be slow, and patients’ responses may be characterized by sound errors. In terms of sentence comprehension, although patients with nonfluent/agrammatic variant often have difficulty with syntactically complex sentence constructions, their understanding of shorter sentences is generally spared. Finally, an additional area of preservation is single-word knowledge, which is typically intact on measures that do not require speech production (i.e., when presented with a single word, the patient can point to the picture that accurately captures the meaning).

### **Non-Language Cognitive Functions in Nonfluent/Agrammatic Variant PPA**

Similar to patients with logopenic variant, nonfluent/agrammatic variant patients’ neuropsychological profile extends beyond language/speech deficits and includes a pattern of performance that may help isolate their diagnosis. These patients typically display mild to moderate difficulty on tasks of executive functions (Libon et al., 2007). Specifically, on tests of verbal fluency, patients with nonfluent/agrammatic variant typically have more trouble with phonemic than semantic generation. This stems from the recruitment of left frontal networks during phonemic fluency tasks (i.e., generating words that begin with a specified letter). Non-verbal design fluency, with its reliance on predominantly right frontal systems more than left, may be relatively preserved. In nonfluent/agrammatic variant, early involvement of inferior frontal gyrus may also extend to dorsolateral prefrontal regions; thus, patients may have difficulty on tests of set-shifting. It is important to highlight that scores on other executive tasks, such as simple attention and working memory, may *underestimate* functioning in these patients if the tasks have significant verbal demands (e.g., requiring rapid repetition or manipulation of number sequences). Abstract thinking may also be mildly affected, though this cognitive sub-domain is difficult to assess given the heavy verbal requirements of the task.

In other cognitive domains, patients with nonfluent/agrammatic variant may do relatively well, with a few minor caveats. Although episodic memory may be quite good, they may require multiple learning trials when encoding novel verbal information due to (a) slowed speech output and (b) the fact that significant cognitive resources are consumed generating verbal output. Patients may also make speech errors in list-learning recall trials that could result in intrusion errors being scored. Although recall of verbal information may be below average, patients may do quite well on list-learning recognition trials. This pattern suggests that they are clearly able to learn and consolidate new information, but their performance is negatively impacted by verbal output demands on free recall trials. Subtle executive deficits may also result in better recognition than free recall in these patients. Visual episodic memory, in contrast, is typically intact in nonfluent/agrammatic variant PPA.

Visuospatial processing is a cognitive strength in patients with nonfluent/agrammatic variant, which is consistent with the preservation of posterior visual-integration networks. Patients will likely do well on copying complex figures and often will have no trouble with simple mathematical calculations (although complex multiplication problems may pose difficulty due to mild executive dysfunction). Patients’ ability to locate objects in two-dimensional space and to discriminate between faces should be preserved. Consistent with the islands of preservation noted in logopenic variant, if an administrator notices striking impairments in the aforementioned cognitive areas in patients with nonfluent/agrammatic variant, then diagnostic concern is warranted.

### **Semantic Variant**

Semantic variant primary progressive aphasia is a clinical syndrome characterized primarily by a gradual decline in semantic knowledge (Gorno-Tempini et al., 2011; Hodges, Patterson, Oxbury,

& Funnell, 1992; Snowden, Goulding, & Neary, 1989; Wilson et al., 2014). Specifically, patients with semantic variant have anomia and a modal deterioration of single-word comprehension. Their neuropsychological profile is characterized by subtle difficulties with episodic memory and executive functions, and relative preservation of visuospatial skills. The relatively focal language and neuropsychological symptoms stem from neurodegeneration in left hemisphere anterior temporal lobe verbal semantic systems. A disease onset that is centered in the right, rather than left, anterior temporal lobe may result in more notable deficits in person than object recognition as well as prominent changes in behavior and empathy (see Babiak, this issue; Henry et al., 2014). In later stages of the syndrome, the disease extends bilaterally and impacts both anterior temporal lobes; it eventually involves other structures including the amygdala, anterior insula, and orbitofrontal cortex—brain regions that are essential for social regulation and emotion (Sollberger et al., 2009). With progression from the anterior temporal lobe to more distributed brain networks, behavioral changes may become prominent (see additional information on this in the Clinical Observations section).

### **Clinical Observations**

Behaviorally, patients with semantic variant may exhibit changes in social comportment and emotion, and may include features such as disinhibition, elation, coldness, rigidity, apathy, hyper-religiosity, and altered food preferences. In our experience, patients with predominantly right-sided disease may be cold and distant whereas those with predominantly left-sided disease may giggle and smile frequently. These patients often exhibit a dramatic decline in empathy and are impaired in their ability to recognize emotions in others. In patients whose disease begins in the left temporal lobe, social comportment is typically more preserved, but individuals may violate social norms involving personal space and eye contact (Shany-Ur & Rankin, 2011).

### **Speech-Language Functions in Semantic Variant PPA**

Language deficits may be pronounced in semantic variant or may be relatively subtle if caught in the earliest stages. Impairment in object knowledge is the hallmark feature of semantic variant; although patients may ask questions that indicate obvious semantic loss (e.g., “What is a hat?”), those with milder deficits may still maintain weakened semantic networks that enable them to name objects correctly when given multiple choice. In conversational speech, patients with semantic variant may be hypervocal and fluent but exhibit word-finding difficulties and semantic paraphasias. This is consistent with their preserved, if not enhanced, dorsal language network which is responsible for speech production and fluency. Their speech may be circumlocutory and, lacking nouns, may be empty in content.

On formal language testing, semantic variant patients show significant difficulties on tests of confrontation naming or object knowledge. Although an early manifestation of semantic variant may be impairment in comprehension of low-frequency words and objects, patients exhibit increasing difficulty with superordinate semantic categories as the disease progresses. In patients with significant semantic loss, semantic cues may not facilitate confrontation naming. Naming tests that use socioemotional stimuli (e.g., photographs of famous individuals) may be more sensitive to right anterior temporal lobe loss than tests that include images of non-social objects. In terms of reading and writing, patients with semantic variant may make errors associated with surface dyslexia and dysgraphia by regularizing irregular words (e.g., phonetically sounding out irregular words such as “yacht”, which typically require semantic knowledge and whole-word reading to accurately read). Areas of preservation include motor speech and repetition; thus, striking impairment in either of these areas should raise diagnostic concern.

### **Non-Language Cognitive Functions in Semantic Variant PPA**

In addition to language deficits, neuropsychological testing may also reveal other areas of cognitive impairment in semantic variant PPA. On tests of episodic memory, patients with semantic variant may have particular difficulty with the verbal domain. Although it is possible that patients have medial temporal lobe atrophy that interferes with their ability to encode new verbal material, it is likely that impoverished semantic knowledge makes learning a list of verbal material more

difficult than it is for people who have rich contextual associations with the items. In more advanced stages of the disease, poor semantic knowledge may render a typical verbal list-learning task the equivalent of learning a list of nonsense words. Recognition testing may also be poor if the foils are words that lack meaning for the patient. Although there should be preservation of visual episodic memory early in the course, at later stages poor comprehension may negatively impact a patient's performance on tests of visual recall or recognition.

On tests of executive functioning, patients with semantic variant may have early preservation or subtle deficits in some areas. As with memory testing, semantic deficits can interfere with a patient's ability to comprehend complicated test instructions or stimuli. On tests of verbal fluency, patients may have deficits in both semantic and phonemic fluency but have relative preservation in non-verbal design fluency tasks. Higher rates of repetition errors on design fluency may occur if there is atrophy in lateral orbitofrontal cortex, an area that is often affected in later stages. Patients also may have trouble with tests of abstract thinking due to impaired semantic knowledge and an inability to integrate higher-order conceptual constructs. Other areas of executive functioning such as set-shifting, working memory, and processing speed may be intact or only mildly impaired in the early stages of semantic variant PPA. Patients may do very well on tests of verbal color-word response inhibition tests (e.g., Stroop interference) because the words carry reduced salience and thus do not interfere with performance as much as they do in healthy individuals.

Visuospatial functioning is an area of cognition that is typically preserved in semantic variant. Mathematical calculations may be preserved, though patients may need to be reminded about the significance of which operation the sign signifies (i.e., reduced knowledge of arithmetic facts). Patients are able to copy even complicated figures, ascertain the location of objects in two-dimensional space, and determine whether objects are similar or different. Some patients may be very exact in their approach to figure copies and even request to use a straight-edge (e.g., ruler) to make their lines perfectly straight. Notably, there is emerging evidence that visuospatial processing may even be enhanced in these patients (Z. A. Miller & Miller, 2013). Heightened attention to visual detail has been reported, and patients may develop new artistic talents and interest in visual arts as their disease progresses and they lose language functioning.

## ***Neuropsychological Differentiation of PPA Syndromes***

Formal neuropsychological testing helps corroborate individual PPA diagnoses and differentiate syndromes from each other. A summary of cognitive profiles for each PPA syndrome is displayed in Table 1.

Table 1. Cognitive Impairments in PPA Syndrome.

Cognitive Domain	PPA Clinical Syndromes: Degree of Impairments		
	lvPPA (Logopenic Variant)	nfvPPA (Nonfluent Variant)	svPPA (Semantic Variant)
<b>Calculations</b> Numerosity Arithmetic Facts Complex Calculations	XX  XX	  X	  X
<b>Executive Functions</b> Task Switching Echoic and Working Memory Cognitive Control and Inhibition Phonemic and Semantic Fluency	(X) XXX  X	XX X X X	(X)   X
<b>Language and Speech</b> Syntax/Grammar Motor Speech Production Confrontation Naming Fluency Reading Repetition Single Word Comprehension	   XX XX XX XXX	  XX XX X XXX (X) X	   XXX  XX  XXX
<b>Memory Consolidation</b> Verbal/Auditory Memory Visual Memory	XX X	(X)	X X
<b>Visuospatial Functions</b> Visual Localization Visuoconstruction Face Perception	X X		(X)
Displays the neuropsychological impairments noted in each PPA syndrome, grouped by cognitive domain. Level of impairment is denoted by an 'X', with 'XXX' suggesting severe impairment and likely a cardinal cognitive feature, 'XX' implying mild to moderate impairment, and 'X' signifying subtle to mild impairment. Parenthetical indicators [e.g. (X)] suggest that subtle difficulties may be noted, or are inconsistently evident in these patients.			

The cognitive phenotype of patients with logopenic variant diverge from nonfluent/agrammatic variant in multiple ways, including their relative preservation of motor speech; thus while patients with logopenic variant may make phonological paraphasias in speech, frank sound distortions are less common (although not entirely absent) than in nonfluent/agrammatic patients. This is consistent with the underlying neuroanatomy of logopenic variant, as these patients do not evidence pathological changes in areas important for motor programming (i.e., inferior motor strip). Patients with logopenic variant also evidence sparing of basic syntactic structure, and can typically generate more words per minute than patients with nonfluent/agrammatic variant (Ash et al., 2013). Relative to semantic variant patients, logopenic variant patients display *slower* speech rates and are more likely to generate phonological paraphasias. Preliminary evidence suggests they may have less difficulty accessing nouns in speech, and perform better than semantic patients on a single word comprehension task. A specific emphasis on “single word” comprehension is integral to this differentiation, as logopenic variant patients will begin to show a breakdown in comprehension consistent with their phonological working memory impairment when presented with multiple words.

In terms of non-language neuropsychological presentations, logopenic variant patients display disproportionate impairment on measures of numerosity and calculations relative to semantic and nonfluent/agrammatic variants, although semantic variant patients may have difficulty retrieving arithmetic facts. In contrast to logopenic variant patients, it is rare for a semantic or nonfluent/agrammatic patient to have difficulty with echoic memory or visuospatial functions, which is consistent with their anterior temporal and inferior frontal anatomy, respectively. Furthermore, striking executive dysfunction is much more common in nonfluent/agrammatic patients and, to a lesser extent, logopenic patients than individuals with semantic variant. As summarized previously, memory functions are deleteriously affected in logopenic relative to nonfluent/agrammatic variant patients, with semantic patients showing artificially low performance due to their loss of semantic knowledge.

## Conclusion

In summary, the three PPA syndromes are characterized by specific cognitive phenotypes that extend beyond prototypical speech-language impairments. The diagnostic criteria for each syndrome understandably highlight speech-language symptoms and further mandate an early prominence of these cardinal features. Broad neural networks that support both language and non-language functions are affected in PPA syndromes, however. Thus a comprehensive assessment of additional neuropsychological domains may aid in solidifying and subtyping PPA diagnoses. We acknowledge that although research on neuropsychological features of PPA remains in a relatively early stage, through a careful assessment of language, non-language, and behavioral symptoms, a comprehensive understanding of the clinical presentation emerges and allows the clinician to appropriately establish differential diagnoses and isolate the specific PPA syndrome.

## References

- Ash, S., Evans, E., O'Shea, J., Powers, J., Boller, A., Weinberg, D., . . . Grossman, M. (2013). Differentiating primary progressive aphasias in a brief sample of connected speech. *Neurology*, *81*(4), 329–336.
- Baddeley, A. (2003). Working memory: looking back and looking forward. *Nat Rev Neurosci*, *4*(10), 829–839.
- Banks, S. J., & Weintraub, S. (2008). Neuropsychiatric symptoms in behavioral variant frontotemporal dementia and primary progressive aphasia. *Journal of Geriatric Psychiatry and Neurology*, *21*(2), 133–141.
- Gorno-Tempini, M. L., Brambati, S. M., Ginex, V., Ogar, J., Dronkers, N. F., Marcone, A., . . . Miller, B. L. (2008). The logopenic/phonological variant of primary progressive aphasia. *Neurology*, *71*(16), 1227–1234.
- Gorno-Tempini, M. L., Hillis, A. E., Weintraub, S., Kertesz, A., Mendez, M., Cappa, S. F., . . . Grossman, M. (2011). Classification of primary progressive aphasia and its variants. *Neurology*, *76*(11), 1006–1014.
- Grossman, M., Mickanin, J., Onishi, K., Hughes, E., D'Esposito, M., Ding, X. S., . . . Reivich, M. (1996). Progressive nonfluent aphasia: language, cognitive, and PET measures contrasted with probable Alzheimer's disease. *Journal of Cognitive Neuroscience*, *8*(2), 135–154.
- Grossman, M. (2012). The non-fluent/agrammatic variant of primary progressive aphasia. *Lancet Neurology*, *11*(6), 545–55.
- Henry, M. L., & Gorno-Tempini, M. L. (2010). The logopenic variant of primary progressive aphasia. *Current Opinion in Neurology*, *23*(6), 633–637.
- Henry, M. L., Wilson, S. M., Ogar, J. M., Sidhu, M. S., Rankin, K. P., Cattaruzza, T., . . . Seeley, W. W. (2014). Neuropsychological, behavioral, and anatomical evolution in right temporal variant frontotemporal dementia: A longitudinal and post-mortem single case analysis. *Neurocase*, *20*(1), 100–109.
- Hodges, J. R., Patterson, K., Oxbury, S., & Funnell, E. (1992). Semantic dementia. Progressive fluent aphasia with temporal lobe atrophy. *Brain*, *115*(Pt 6), 1783–806.
- Libon, D. J., Xie, S. X., Moore, P., Farmer, J., Antani, S., McCawley, G., . . . Grossman, M. (2007). Patterns of neuropsychological impairment in frontotemporal dementia. *Neurology*, *68*(5), 369–375.
- Mesulam, M. M. (1982). Slowly progressive aphasia without generalized dementia. *Annals of Neurology*, *11*, 592–598.



- Mesulam, M. M., Wieneke, C., Thompson, C., Rogalski, E., & Weintraub, S. (2012). Quantitative classification of primary progressive aphasia at early and mild impairment stages. *Brain*, *135*(Pt 5), 1537–1553.
- Miller, Z. A., & Miller, B. L. (2013). Artistic creativity and dementia. *Progress in Brain Research*, *204*, 99–112.
- Rogalski, E., Cobia, D., Harrison, T. M., Wieneke, C., Weintraub, S., & Mesulam, M. M. (2011). Progression of language decline and cortical atrophy in subtypes of primary progressive aphasia. *Neurology*, *76*(21), 1804–1810.
- Rohrer, J. D., & Warren, J. D. (2010). Phenomenology and anatomy of abnormal behaviours in primary progressive aphasia. *Journal of the Neurological Sciences*, *293*(1–2), 1752–1756.
- Shany-Ur, T., & Rankin, K. P. (2011). Personality and social cognition in neurodegenerative disease. *Current Opinion in Neurology*, *24*(6), 550–555.
- Snowden, J. S., Goulding, P. J., & Neary, D. (1989). Semantic dementia: a form of circumscribed atrophy. *Behavioral Neurology*, *2*, 167–182.
- Sollberger, M., Stanley, C. M., Wilson, S. M., Gyurak, A., Beckman, V., Growdon, M., . . . Rankin, K. P. (2009). Neural basis of interpersonal traits in neurodegenerative diseases. *Neuropsychologia*, *47*(13), 2812–2827.
- Wilson, S. M., Henry, M. L., Besbris, M., Ogar, J. M., Dronkers, N. F., Jarrold, W., . . . Gorno-Tempini, M. L. (2010). Connected speech production in three variants of primary progressive aphasia. *Brain*, *133*(Pt 7), 2069–2088.
- Wilson, S. M., DeMarco, A. T., Henry, M. L., Gesierich, B., Babiak, M., Mandelli, M. L., . . . Gorno-Tempini, M. L. (2014). What role does the anterior temporal lobe play in sentence-level processing? Neural correlates of syntactic processing in semantic variant primary progressive aphasia. *Journal of Cognitive Neuroscience*, *26*(5), 970–985.