# Dementia

# Assessment of Language Function in Dementia

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Impairment in language is a common finding among individuals with dementia and can be a presenting symptom, particularly in Alzheimer's dementia and primary progressive aphasia. Early recognition of language dysfunction can help with an accurate diagnosis, management, and prognosis. There are numerous established and validated language evaluation protocols. This article provides a simple means for the primary care physician to identify and evaluate language disorders in dementia, but it is not meant to replace established protocols.

*Key words:* aphasia, dementia, primary progressive aphasia, semantic dementia, *Alzheimer's disease* 

## Introduction

Language is the ability to encode ideas into words or symbols for communication to someone else<sup>1</sup> and involves speaking, comprehending, reading, and writing. Language should be differentiated from speech, which is the motor aspect of spoken language production.<sup>2</sup> Impairments in speech arise from any process that disrupts the neuraxis from the cortex to muscle and encompass dysarthria (disturbance in articulation) and dysphonia (disturbance in the production of vocal sounds).

Aphasia is an acquired language impairment caused by damage to the brain. It may involve language production, comprehension, or both. Although stroke is the most common cause of aphasia seen in general practice, progressive aphasias can occur in different dementing syndromes. The aphasic syndromes caused by stroke (outlined in Figure 1) bear a resemblance to those associated with neurodegenerative disease, but the terms used to describe stroke aphasia are not generally used to describe language impairments in dementia (with the possible exception of *transcortical sensory aphasia*). In addition, the brain areas affected are usually more localized in aphasia caused by stroke than in dementia.<sup>3</sup>

Identification of language impairment is important in dementia because it aids in the accurate diagnosis of a specific type of dementia, alters the prognosis, and changes the management. The language impairment can be missed this frequently leads to misdiagnosis.<sup>4</sup>

# Types of Aphasia Commonly Seen in Dementia

The two main dementia syndromes in which language impairment is likely to be an early feature are Alzheimer's dementia and primary progressive aphasia. In Alzheimer's dementia, the cognitive impairment extends beyond language and typically involves episodic (i.e., anterograde or day-to-day) memory. In primary progressive aphasia, gradual deterioration in language skills occurs in the context of relatively preserved nonverbal skills and activities of daily living.<sup>5,6</sup> With progression, severe communication difficulties develop and more generalized cognitive impairment usually appears.

The type of aphasia seen in Alzheimer's dementia is dependent on the stage of the disorder (Table 1). In the early stages, there may be mild wordfinding difficulties with occasional semantic paraphasias (i.e., semantic substitutions, such as saying aunt instead of sister), but speech remains fluent and grammatically correct. This is known as anomic aphasia. With progression, these individuals exhibit transcortical sensory aphasia, in which there is clear anomia and comprehension is affected.<sup>7</sup> In the moderate to severe stages of Alzheimer's, there is a loss of fluency, increased paraphasias (use of incorrect words as well as incorrect pronunciation), and poor comprehension. There is a reduction in output, and in the severe stages, there may be echolalia (repetition of words or phrases said by somebody else) and verbal stereotypies (repetition of meaningless words or phrases).8

The primary progressive aphasias are classified as either fluent or nonfluent. In the fluent variant, speech remains fluent, with normal prosody, and is wellarticulated and grammatically correct yet becomes progressively circumlocutory and lacking in content words.9 The language impairment is associated with a degradation of semantic memory and, therefore, the fluent variant is often referred to as semantic dementia. In the nonfluent variant, speech is effortful, hesitant, and faltering, with phonemic paraphasias (for example, *rhinoceros* becomes rhinosus).<sup>10</sup> The characteristics of the aphasia in primary progressive aphasia and in Alzheimer's dementia are summarized in Table 1.

# **Obtaining a History**

The elements to obtain in the history are no different from those taken in a history of a person with typical Alzheimer's dementia and include "(i) symptoms at onset, (ii) tempo of evolution of symptoms, (iii) impact on work and family life, (iv) issues of safety, (v) family history of dementia, (vi) presence of cerebrovascular risk factors, and (vii) past medical history."<sup>11</sup> In addition to the patient's account, it is important to obtain a corroborating history from a relative or caregiver and to inquire about any changes in the patient's spoken language production and comprehension, as well as reading and writing.

Any complaint of memory problems should be queried closely. Patients and their caregivers often describe any major cognitive problem as a "memory problem," but they are not necessarily referring to the anterograde memory loss seen in Alzheimer's dementia. Examples of the memory loss should be requested as they may indicate primary language impairment. For example, patients may forget the name of something or a word's meaning (e.g., asking, "What is asparagus?"), indicating a loss of linguistic and/or semantic knowledge, rather than a memory problem.

#### Language Assessment

The Mini-Mental State Examination (MMSE)<sup>12</sup> does allow some formal testing of language, but additional detailed testing is required to gain insight into the type of underlying dementia. Listening carefully to a patient's spontaneous speech is a key aspect of the language assessment (Table 2). This can be done by asking open-ended questions such as, "Why have you come to see me?" and by asking the patient to describe a complex picture, such as the well known "cookie theft" picture from the Boston Diagnostic Aphasia Examination.<sup>13</sup> In a pinch, a picture from a magazine or newspaper could be used. When listening to spontaneous speech, the clinician should pay attention to articulation (clarity or distortion of speech), fluency (rate of speech, phrase length), grammatical accuracy and variety, word finding (Are there pauses to search for words? Is a good range of vocabulary used?), and prosody or melodic line.<sup>14</sup>

Naming (Table 3) is tested by asking the patient to name objects around the room, body parts, or line drawings, such as those in the Boston Naming Test.<sup>15</sup> Naming is typically impaired in dementia, and the types of errors can be informative. Phonemic or semantic errors can be observed and suggest nonfluent or fluent progressive aphasia, respectively. Patients who describe what a word means instead of providing the name are employing circumlocution. When patients fail to recognize what an object is, they are exhibiting visual agnosia caused by the loss of

Table 1: Summary of Language Impairments in Alzheimer's Dementia and Primary Progressive Aphasia						
	Alzheimer's Dementia		Primary Progressive Aphasias			
	Early Stage	Moderate-Severe	Nonfluent Variant	Semant	tic Dementia Variant	
Spontaneous speech	Fluent, grammatical	Nonfluent, echolalic, neologisms	Effortful, short phrase length, obvious word- finding difficulty	correct	melodic, grammatically , obvious word- difficulty	
Paraphasic errors	Semantic	Semantic and phonemic	Phonemic	Semantic		
Repetition	Intact	Impaired	Impaired	Intact (for single word)		
Naming	Impaired (mild)	Impaired	Impaired	Impaired		
Comprehension of words	Intact	Impaired	Intact	Impaired		
Syntactic comprehension	Intact	Impaired	Impaired	Intact		
Reading	± Intact	Impaired	Impaired	Impaired (surface dyslexia)		
Writing	± Intact	Impaired	Impaired	Impaired (surface dysgraphia)		
Word and object knowledge (semantic knowledge)	Intact for more frequently used words and objects; impaired for less frequently used words and objects	Impaired	Intact	Impaired		
		Frontotemporal dementia				
		Progressive supranuclear palsy Alzheimer's disease				
			Alzheimer's disease			

#### Assessment of Language Function in Dementia

semantic knowledge for the object. Circumlocution and visual agnosia can be observed in both early Alzheimer's dementia and semantic dementia; initially, this occurs with infrequently encountered words or objects, but with disease progression, it happens even with (previously) familiar words or objects.

Repetition of words and sentences (see Table 3) should also be tested as this can differentiate the types of aphasia seen in dementia. It also provides localizing value: impaired repetition implies a lesion in the perisylvian area, while intact repetition in conjunction with aphasia implies a lesion in the extraperisylvian area and signifies a transcortical aphasic disorder (see Figure 1).

Comprehension is tested by asking the patient to follow a series of one-, two-, and three-step commands (see Table 3). Additional tests in comprehension can also include asking the definition of words to test the patient's semantic knowledge or asking for an interpretation of a story by providing a short scenario followed by a question. An example of story interpretation is, "A tiger and lion fought in the jungle. The tiger *was eaten by* the lion. Who won?"

Reading comprehension (see Table 3) can be tested by having the patient read commands and follow them—similar to one of the items in the MMSE. Reading aloud can be tested by having the patient read aloud either a standardized paragraph or one from a magazine. Reading of single words should be tested if paragraph reading is not successful.

Writing (see Table 3) can be tested by asking the patient to write a sentence. The sentence from the MMSE should suffice.

	Table 2: What to Look for in Assessment of Spontaneous Speech						
Language Domain	Description	Related Dementia Syndrome	Examples				
Fluency	Dysfluent speech hesitant and faltering, with abnormal prosody and reduced phrase length	Primary progressive aphasia— nonfluent variant	<ol> <li>"My speech I can't tell the, I can't express it."</li> <li>"I, ah, umm I feel I forget it, ah, yes."</li> </ol>				
Fluency	Fluent speech normal in rate, phrase length, and prosody	Alzheimer's dementia Semantic dementia					
Naming or word finding	Anomic speech is empty, lacking meaning, and reduced in content words; may be pauses while the patient searches for a word; often is substitution of generic or superordinate terms (e.g., <i>thing, animal</i> ), as well as the use of circumlocution	Semantic dementia Primary progressive aphasia—nonfluent variant Alzheimer's dementia (naming impairment will be mild in the early stages)	Empty speech: 1. "You can see out there and the things are out there." 2. "If you are reading and writing, and then just carrying on with the story, then the next step is I'm not sure." Circumlocution: 1. "Something that goes up in the air." (to indicate <i>helicopter</i> ) 2. "You can turn things with it." (to indicate <i>screwdriver</i> — note also the use of the general term <i>thing</i> )				
Grammar	Agrammatism—may be grammatical errors, and speech is reduced in grammatical complexity	Agrammatic production both reported and disputed in primary progressive aphasia— nonfluent variant; it is however accepted that there is grammatical comprehension impairment	<ol> <li>"My wife, umm, teacher, umm, full- time, umm, umm, children, umm, school."</li> <li>"I did four cars and and yes  four cars, but not very good. One don't like it, but two not bad."</li> </ol>				
Paraphasic errors	Phonemic (literal)— substitution of one sound for another	Primary progressive aphasia— nonfluent variant	<i>Ticycle</i> instead of <i>bicycle</i> <i>Elefisis</i> instead of <i>elephant</i>				
	Semantic (verbal)— substitution of a word that is semantically related to the target	Semantic dementia	<i>Car</i> instead of <i>truck</i> <i>Hippopotamus</i> instead of <i>rhinoceros</i>				

#### Table 2: What to Look for in Assessment of Spontaneous Speech

The sentence should be examined for grammar, spelling errors, phrase length, and punctuation. In addition, writing to dictation (of regular and exception words) and copying should be tested if spontaneous writing is not successful. Patients with a progressive aphasia often have insight into their difficulties and are frustrated by their problems with communication. These difficulties can be exacerbated when they are asked to converse or to perform tests quickly. Extra

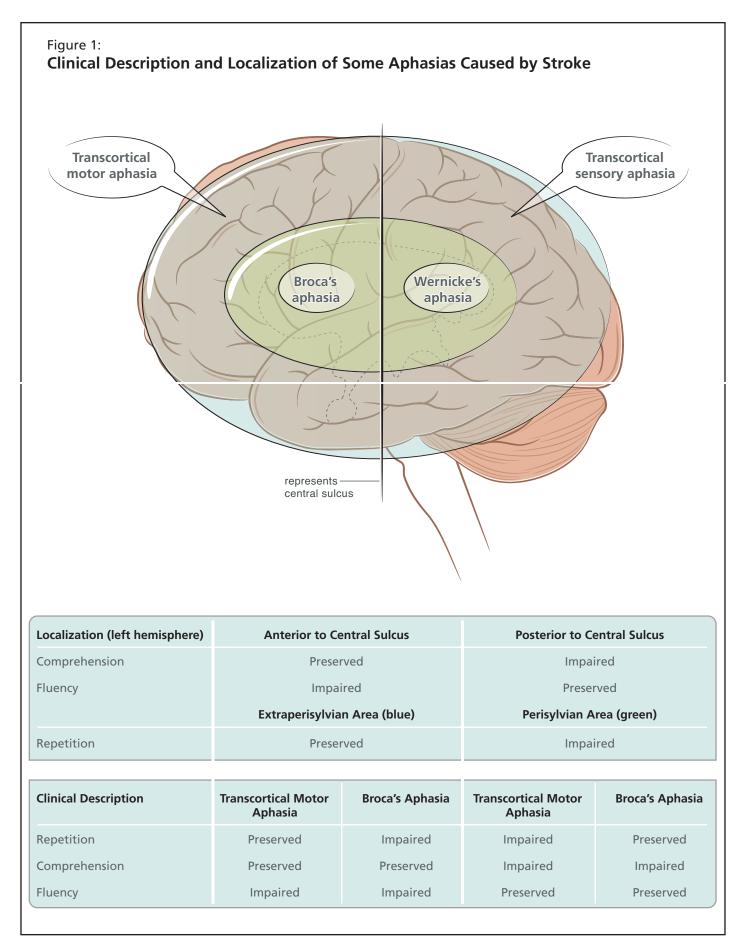
time must be provided to allow these patients to communicate.

# Putting It All Together: A Case Example

A 66-year-old, right-handed woman with

Language Domain	Examples of Assessment Methods		
Naming	Ask the patient to name items around the room or on your person (e.g., watch—including the crystal, stem, band; tie; collar; shoe—including eyelet, sole, heel) Ask the patient to name the pictures on the 15-item CERAD naming <sup>23</sup>		
Repetition of sentences	<ul> <li>Ask the patient to repeat unfamiliar sentences/phrases; examples:</li> <li>"No ifs, ands, or buts"</li> <li>"Go into the garden and pick up the beans."</li> <li>"The judge sentenced the criminal."</li> <li>"The prime minister lives in Ottawa."</li> </ul>		
Comprehension (auditory)	<ul> <li>Ask the patient to perform (in the absence of apraxia):</li> <li>Single one-step commands (e.g., "Touch your nose" or "Point to the door")</li> <li>Followed by two-step commands (e.g., "Touch your nose and then point to the floor")</li> <li>Followed by three-step commands (e.g., "Take this piece of paper in your left hand, fold it in half, and put it on the floor")</li> <li>Can give grammatically complex commands (e.g., "Before touching your nose, point to the floor")</li> </ul>		
Comprehension (semantic knowledge)	<ul> <li>Ask the patient to define words or perform tasks requiring decoding of semantic knowledge; examples:</li> <li>"Point to a source of illumination."</li> <li>"Point to the surface that you walk on."</li> <li>"Define island." "Define pyramid."</li> </ul>		
Reading	<ul> <li>Ask the patient to read the following commands (examples) and perform them:</li> <li>"Close your eyes."</li> <li>"Touch your nose."</li> <li>"Before touching your chin, point to the ceiling."</li> <li>"Open your mouth after you point to your chair and the floor."</li> <li>Ask the patient to read a paragraph from the newspaper or magazine—observe for reading effort, pronunciation of words, melody/cadence</li> </ul>		
Writing	<ul> <li>Ask the patient to write a sentence to examine for grammar, spelling errors, phrase length, and punctuation; examples:</li> <li>Why the patient came to see you</li> <li>A description of the weather or a picture shown to them</li> <li>Ask the patient to write a sentence to dictation (e.g., "Some water is not good to drink.")</li> <li>Ask the patient to copy a sentence</li> </ul>		

#### Table 3: Formal Language Assessment

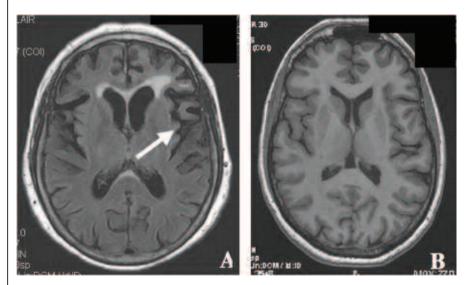


10 years of formal education was assessed for the insidious onset of wordfinding difficulties of 2 years' duration. As a result of her difficulties, she would sometimes stop in mid-sentence and be unable to continue, or would produce the beginning of a word but not finish it (e.g., rec— for *recognize*). Her sentences had become shorter in length, and she had been making spelling errors on simple words. She had not been disinhibited.

Cognitive testing revealed a score of 18 out of 38 on the Short Test of Mental Status<sup>16</sup>—a bedside test that is similar to the MMSE-with sparing of her memory and visuospatial and calculation skills. Examination of her spontaneous speech revealed effortful nonfluent speech, phonemic (e.g., *jusee* for *judge*) more than semantic paraphasic errors (e.g., pies instead of muffins), and the use of pantomime for words she knew but could not say (e.g., she mimed sewing manoeuvres but could not say the word). She had impaired repetition of sentences but not single words, impaired comprehension (she could not follow two-step commands), impaired naming (with 16 out of 30 on the Boston Naming Test) and could not write a sentence. She wrote, "I live at 75, 1203 in—" in an attempt to write down her address. Her neurological examination was normal.

A review of this case illustrates several characteristics of a progressive aphasia. The history shows that this person's problem started insidiously and is characterized by nonfluent speech, spelling mistakes, and anomia. The language assessment not only confirms the problems seen in the history but also demonstrates paraphasic errors and impaired comprehension, writing, and naming. Despite her aphasia, she tested well in her other cognitive domains. The language and cognitive assessment localizes her difficulties to the perisylvian area of her left hemisphere. Indeed, magnetic resonance images of her brain revealed focal atrophy in this area (Figure 2). Her presentation is typical of a patient with the nonfluent variant of primary progressive aphasia.

#### Figure 2: Axial Magnetic Resonance Imaging Brain Scans



Panel A: Axial MRI scan of the brain of a patient with nonfluent primary progressive aphasia, demonstrating bilateral perisylvian atrophy which is more extensive on the left (arrow); this patient's case is the example described in the text. Panel B: Axial MRI of a normal brain.

Source: Courtesy Dr. David Tang-Wai

# The Evolution of Primary Progressive Aphasias: The Need to Monitor Additional Symptoms

Although a discussion of the evolution of symptoms in primary progressive aphasia is beyond the scope of this article, it is important to note that the aphasia is only the *presenting* symptom. It is expected that the language disorder will progressively deteriorate and that, unfortunately,

additional features will develop. These features will further define the actual underlying disorder (see Table 1). The family physician ought to be aware of this possibility and inquire about (1) symptoms suggestive of changes in personality or behaviour consistent with either apathy or disinhibition—indicative of frontotemporal dementia or Pick's disease; (2) asymmetric parkinsonism associated with ideomotor apraxia indicative of corticobasal degeneration;

#### **Key Points**

Language dysfunction (in the absence of memory impairment) can be a presenting feature of a degenerative dementia.

A brief language assessment can identify the language issues.

Aphasia may be a feature in Alzheimer's dementia, as well as in primary progressive aphasia.

Evaluation of the nature of the language impairment can be informative in staging of Alzheimer's disease and in identification of the variant (fluent versus nonfluent) of primary progressive aphasia.

When a progressive language disorder is identified, follow-up is essential as further neurological signs and symptoms may develop; the nature of these features depends upon the underlying disorder.

(3) axial rigidity, supranuclear gaze palsy, and falls—indicative of progressive supranuclear palsy; (4) prominent anterograde memory impairment, acalculia, and visuospatial impairment—indicative of Alzheimer's disease; and (5) dysphagia, falls, fasciculations, and muscle weakness—indicative of motor neuron disease.<sup>17–21</sup> A neurological examination must be performed and, as a minimum, the following should be investigated: fasciculations, muscle atrophy, rigidity or spasticity, bulbar and/or limb weakness, hyperreflexia, and shuffling or spastic gait.<sup>22</sup>

# Conclusion

Accurate identification of language impairment will improve diagnosis and management of progressive aphasia. It is therefore important to evaluate language functioning in early dementia. Careful listening to the patient's speech and a brief language assessment can provide key information. If language impairments are identified in the context of dementia, the dementia is most likely to be Alzheimer's dementia or primary progressive aphasia. More detailed language assessment and neurological evaluation can be obtained via referral to a memory clinic. ga

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