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Chapter

Primary Progressive Aphasia (PPA)

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Abstract

Primary progressive aphasia (PPA) is a rare neurological condition that affects an individual’s ability to communicate. PPA is a syndrome characterized by a ‘progressive worsening of language with preservation of the activities of daily living and evidence of relatively normal non-verbal abilities on neuropsychological testing’. It commonly appears initially as a disorder of speaking (an articulatory problem) progressing to nearly total inability to speak in its most severe stage, while comprehension remains relatively preserved. This chapter provides an overview of the characteristic features of PPA, its classification, assessment, and rehabilitation options.

Keywords: Primary Progressive Aphasia, neurology, speech-language, dementia, assessment, management

1. Introduction

Primary Progressive Aphasia (PPA) is a type of dementia characterized by a loss in one or more language functions in people under the age of 65 (and occasionally as early as the 40’s). It is a type of dementia and the condition is caused due to degeneration of nerve cells in the brain which control our ability to communicate [1].

PPA is a condition marked by a steady deterioration of language skills while maintaining everyday activities and showing relatively normal nonverbal abilities in neuropsychological testing. It frequently begins as a speech deficit (an articulatory problem), advancing to near total inability to speak in its most severe stage, but understanding remains largely intact.

Other neurological illnesses, on the other hand, exist in which language degeneration is simply one aspect of a broader, progressive decrease in mental processes such as memory, attention, visuospatial skills, reasoning, and the ability to perform complicated physical activities. The generation of phonemic paraphasias in naming was offered as a further criterion for separating PPA from progressive aphasia in likely Alzheimer’s disease [2].

2. A retrospective delineation

Pick [3] described a patient who had a progressive language disorder that was linked to left temporal lobe atrophy. This seminal paper was followed by a flood of cases [1, 2], and it became clear that PPA can present in both fluent and non-fluent people. Over the next few decades, there were intermittent incidents [1, 4].
Mesulam's describes PPA in the context of atrophy seen in the left focal perisylvian region or temporal lobe [5]. Speech is grammatically structured and well-articulated but gradually loses content words.

The study of patients with a progressive deficit in semantic memory began at the same time as the study of cognitive issues linked with neurodegenerative disease [6, 7]. The rising anomia exhibited in these people, according to Warrington, was caused by a fundamental loss of semantic memory, which impaired object recognition and knowledge, as well as word discovery and comprehension. Following more evidence of impaired nonverbal conceptual understanding (semantic memory) as well as anomia in these patients, the term 'SD' was coined. The 1998 consensus declaration is often quoted. Associative agnosia (difficulty recognizing/identifying items) and/or prosopagnosia (difficulty recognizing/identifying known or renowned individuals) were supported as criterion for SD in the widely recognized 1998 consensus statement. PPA is currently included in the frontotemporal lobar degeneration, fronto-temporal dementia, Pick's disease, or Pick's complex family of disorders.

3. Characteristics of PPA

Main characteristics of PPA [8] are as follows:

• Age of onset ranges from 40 to 75, with a mean of 59.3 years.

• The involvement of males predominates over females, with the ratio of 2:1.

• The duration of isolated language symptoms can range from 1 to 15 years with a mean of 5.3 years.

• Autopsy findings reported for 14 cases disclosed: Pick's disease in 4, Creutzfeld—Jakob's disease in 3, Alzheimer's disease in 3, focal spongiform degeneration in 2, and nonspecific cellular changes in 2. The diversity of pathology does not support the existence of a specific disease underlying isolated language decline.

• Of the 47 cases with CT scan, 13 were normal, 5 showed a diffuse abnormality, 10 had greater left than right abnormality, and 19 had left hemisphere abnormality only.

• Most of the reported cases had predominantly fluent, anomic, or Wernicke’s like aphasia, but 12 cases with nonfluent or Broca’s aphasia have been described.

• The description of cases with PPA has aided in the long-running, more general debate over whether a stage model or a subgroup model [9] might be more appropriate for explaining the neuropsychological consequences of certain degenerative brain diseases, as they appeared to support a subgroup model.

• Subgroup models do not presume that cognitive performance is disintegrated uniformly. Rather, they assume that observed disparities between patients and patient groups are qualitative in origin and represent impairments in various cognitive modules.

• Stage models, on the other hand, assume a global progression of cognitive deficiencies with solely quantitative changes over time, with observed disparities.
between patients and patient groups explained by distinct points on a one-dimensional time-axis.

**Early symptoms:**

- Difficulties recalling the names of people and objects,
- Difficulties expressing one's thoughts orally and sometimes difficulty understanding others.
- Memory of recent events, attention/concentration, judgment and reasoning abilities, and visuospatial abilities are relatively normal.
- Some patients tend to say or nod “yes” for “no” and vice versa.
- Gradual difficulty in one or more of the following language functions like speaking, understanding, reading and writing.
- Inability to follow conversations or verbal instructions.
- Inability to read or write a letter.
- Decreasing content in speech or garbled speech to eventually becoming mute.

**Later symptoms:**

- As PPA progresses, it becomes increasingly difficult to communicate verbally by any means.
- The capacity to comprehend what others are saying or reading deteriorates as well.
- Inability to understand speech.
- Difficulty following conversations, especially in bigger groups.
- Requesting information to be repeated and misinterpreting what is said despite normal hearing.
- Early in the course of the illness, speech may become empty of any genuine information and difficult to grasp.
- Eventually, speech may be unable to communicate, leading to muteness.
- Math and calculating problems.
- Loss of ability to do even elementary mathematical processes.

**Diagnostic criteria for PPA [10]:**

- A distinctive feature is an insidious onset and slow advancement of word-finding, object naming, or word understanding as shown during spontaneous conversation or as tested through formal neuropsychological language tests.
• For at least 2 years following, the language impairment is responsible for all limitations in daily living activities.

• Premorbid linguistic function is intact (except for developmental dyslexia).

• Within the first 2 years of the illness, there was no notable apathy, disinhibition, forgetfulness for recent events, visuospatial impairment, visual recognition deficiencies, or sensory-motor dysfunction.

• Even in the first 2 years, acalculia and ideomotor apraxia may be present (mild constructional deficits and perseveration are also acceptable as long as neither visuospatial deficits nor disinhibition influence daily living activities).

• Other domains may be impacted after the first 2 years, but the language remains the most impaired function and deteriorates quicker than other afflicted domains throughout the illness.

• Neuroimaging has revealed that there are no “particular” reasons such as stroke or tumor.

4. Types of Primary Progressive Aphasia

The most common type of brain degeneration found after brain autopsy in individuals with PPA (60% of cases) is Nonspecific degeneration (NSD) where there is evidence of brain cell death, but no features of Alzheimer’s or Pick’s disease. Less commonly, Pick or Alzheimer’s disease may be found.

Primary progressive aphasia, or PPA, is a group of Fronto-Temporal Dementia (FTD) symptoms defined by a progressive loss of capacity to talk, read, write, and understand what others are saying. When the following three conditions are met, PPA is diagnosed:

1. There is a steady deterioration in verbal ability (not just speech).

2. At first, the sole disability is a linguistic barrier.

3. A neurological illness is the root reason.

PPA is further divided into three clinical subgroups based on the language abilities that are most impacted, according to experts.

4.1 Semantic variant of PPA

The increasing loss of word meanings is a feature of semantic variant PPA (svPPA). The disorder is sometimes known as semantic dementia if there are extra substantial issues recognizing things or faces. Other language skills remain unchanged, such as the ability to make speech and repeat words and sentences uttered by others. However, while the affected person may continue to speak fluently, their speech becomes hazy and difficult to comprehend due to the omission or substitution of several words. People with svPPA (also known as PPA-S) may demonstrate changes in behavior compared to those seen in behavioral variant of Fronto-Temporal Dementia (bvFTD), such as disinhibition and rigid food preferences, as the disorder progresses.
Signs and symptoms:

- **Anomia**—An inability to recall the names of objects; difficulty “finding the right word.” (“car” for “truck.”)

- **Reduced single-word comprehension**—The person affected is unable to recall what words mean, especially words that are less familiar or less frequently used.

- **Impaired object knowledge**—Being unable to remember what a familiar object is or how it is used.

- **Surface dyslexia/dysgraphia**—Reading and writing words that do not follow standard pronunciation or spelling conventions can be difficult; such words are spelled or spoken “as if” they do. For example, the person might write “no” instead of “know” or misread “wide” as “broad.”

### 4.2 Non-fluent variant of PPA

People with the non-fluent/agrammatic form of PPA (nfvPPA, also known as PPA-G), also known as progressive non-fluent aphasia or PNFA, have trouble speaking but, can remember the meanings of individual words. Degeneration of the areas of the brain that regulate specific linked muscles causes them to lose their ability to make sounds with their lips and tongue; nevertheless, the muscles themselves are intact. Apraxia of speech is the technical term for these issues (AOS). As a result, their speech becomes labored and slow, and they may appear to be physically struggling to speak. However, speech issues alone are not enough to diagnose PPA. When speech rather than language is the primary difficulty, the diagnosis is progressive AOS rather than PPA. The impairment of grammar is a distinctive characteristic. When speaking, people with nfvPPA make a variety of errors, such as deleting small grammatical terms, wrongly employing word ends and verb tenses, and/or messing up word order in phrases. Some people may eventually develop swallowing difficulties as well as more generalized motor symptoms, comparable to those found in movement-predominant FTDs such as corticobasal syndrome.

**Signs and symptoms:**

- **Apraxia**—difficulty making the necessary lip and tongue movements for speech. This causes distorted or inaccurate speech sounds, as well as slow, strained speech and groping facial and lip motions in an attempt to generate the correct sound. The initial symptom is generally labored speaking. Words with several syllables are the most difficult to create.

- **Agrammatism**—due to omissions and inaccuracies, speech becomes limited to short, simple phrases that are difficult to understand by the listener.

- **Difficulty understanding long or grammatically difficult sentences.** Single-word comprehension is unchanged, but the capacity to grasp long or grammatically difficult sentences is impaired. People with PPA may find it more difficult to comprehend what they see as “too much” verbal information, such as watching television or following a group conversation.

- **Mutism**
• Difficulty swallowing. This occurs later in the disease’s course.

• Movement impairments similar to Parkinson’s disease can arise. Slow, stiff movement, loss of balance or falling easily, trouble moving an arm or leg, and restricted up-and-down eye movement are all possible symptoms.

4.3 Logopenic variant of PPA

When speaking, people with logopenic variant PPA (lvPPA, also known as PPA-L) have trouble choosing words. As a result, individuals may speak slowly and pause frequently while searching for the appropriate word. They can still remember the meanings of words, unlike persons with semantic variant PPA. Speech can be absolutely fluent during the small discussion but become hesitant and halting when the person has to be particular or use a more unfamiliar word, unlike persons with agrammatic PPA. Speech is normally unforced and undistorted. The lvPPA type also has a short attention span for words, which makes it difficult to repeat phrases and sentences. As the condition advances, those who are affected may have difficulty understanding complex sentences.

Signs and symptoms:

• Impaired single-word retrieval
  ○ Finding the correct word while speaking is difficult.
  ○ Due to the time required for word recovery, there are pauses and hesitations.
  ○ An extended description (circumlocution) can be used to replace a lost word.

• Impaired repetition—Longer phrases and sentences pose more difficulty.

• Phonological speech errors—Omissions and substitutions in speech sounds are examples of mistakes. For instance, the affected individual may substitute sounds made at the tip of the tongue, such as “t” or “d,” for sounds made near the neck, such as “k” or “g,” as in “tup” instead of “cup” or “dap” instead of “gap.” They can remove final consonants, such as “slee” rather than “sleep.”

• Phonological paraphasias—A non-word with some of the same sounds is substituted for a real word. For example, the affected person might say “tele-phone” instead of “telephone.”

• Poor comprehension of complex sentences—With single-word comprehension spared.

• Difficulty swallowing—This may develop later in the progression of the disease.

5. Speech, language, and communication assessment of PPA

5.1 History

First, a thorough medical history is taken to determine whether a dementia condition, as stated above, exists. Because the patient may be ignorant of the symptoms
(as in the case of memory loss or personality changes) or unable to describe them owing to aphasia, family members or friends are frequently questioned about the patient's behavior.

Case history and interview:

- It is critical that a thorough case history be obtained.
- Document the initial presentation as well as the emergence of any additional symptoms over time.
- Pay attention to characteristics of speech and language. At the time of evaluation, the degree of linguistic versus cognitive or motoric impairment (e.g., word-finding difficulties, agrammatism, phonological errors)
- Nonverbal cognitive deficits that could worsen over time (e.g., episodic memory impairment).
- Atypical behavioral symptoms (e.g., disinhibition, apathy, loss of empathy) that can arise, particularly in semantic variant PPA, or motoric symptomatology (e.g., limb apraxia, Parkinsonism, dysphagia) that can develop, most commonly in nonfluent/agrammatic PPA. If not previously indicated in the medical record, any family history of dementia or other pertinent medical diagnoses (particularly neurodegenerative disorders) should be investigated.
- Disabilities in hearing and/or vision should be documented.
- Whether the person is monolingual, bilingual, or multilingual.

The therapist should enquire about existing functional communication demands and constraints in order to inform therapy planning. It's crucial, for example, to figure out which communication contexts (work, home, community), partners (family, friends, coworkers), and modes (telephone, face-to-face, written) are most significant in the patient's daily life. If possible, include the individual's primary communication partner(s) to establish a thorough and accurate case history.

5.2 Neurological examination

A neurological examination is performed to see if there is evidence of dementia on a simple mental function screening (the mental status assessment) as well as signs of motor or sensory symptoms that indicate various forms of neurological illnesses could be causing dementia. The neurologist may also arrange tests, such as an MRI scan, to rule out the possibility of a stroke or tumor as the cause of the symptoms.

5.3 Brain imaging studies

This can be done with a CAT scan or an MRI scan. Both of these technologies create an image of the brain, allowing any structural abnormalities like a stroke, tumor, or hydrocephalus—all of which can cause dementia-like symptoms—to be recognized.

The CAT scan and MRI scans in the case of degenerative brain disease may reveal “atrophy,” which indicates “shrinkage” of the brain tissue, or they may not
show anything at all, especially in the early stages. In fact, the report is frequently returned as “normal.” However, this just implies that no evidence of a tumor or stroke exists. It is unable to provide information on the microscopic degenerative changes that have occurred.

5.4 Neuropsychological examination

It provides a more thorough assessment of mental health. This is especially crucial in the early stages of sickness, when a standard screening examination may fail to discover the patient’s difficulties. This takes several hours and consists of tests of mental talents, such as attention and focus, language learning and memory, visual perception, logic, and mood, conducted by paper and pencil or computer. The results can reveal whether there are any irregularities in thought or behavior, as well as their severity (mild, moderate, or severe). Because we normally test memory by telling someone something and then asking them to repeat it later, demonstrating that people with PPA have intact memory can be difficult. In an individual with PPA, it may be impossible to repeat back the information because of the aphasia.

As a result, it’s critical that testing is done adequately to ensure that there is not an actual memory loss. To yet, the only way to objectively diagnose dementia is to undergo a neuropsychological evaluation. There are currently no dementia-specific blood tests or other physiological diagnostics available. This is especially helpful if the symptoms are modest or in the early stages.

5.5 Psychiatric evaluation

Sometimes there will also be a need for a psychiatric evaluation. This may be the case when it is not clear if the changes in behavior are due to depression or another psychiatric disturbance.

5.6 Physiological evaluation

PPA impacts not just the person who is afflicted with the disorder, but also those who are close to them. Relationships, the ability to continue working, the ability to execute numerous ordinary tasks, and the ability to convey even the most basic requirements are all affected by the disease. Although there are several resources accessible for people with memory loss, there are far fewer resources available for people with PPA, their relatives, and friends. A consultation with a social professional who is experienced with PPA can address these concerns and offer advice on how to deal with daily frustrations and problems.

5.7 Speech and language evaluation

Because the primary symptom of PPA is a reduction in language abilities, it’s critical to figure out which aspects of language use are most affected, how badly they are affected, and what may be done to improve communication. A speech-language pathologist (SLP) examines various areas of language in-depth and might provide recommendations to improve communication. Family members should be included in treatment sessions to learn how to communicate more effectively.

5.8 Formal assessment

The following tests can be used to assess various functions in patients with PPA:
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• Mini-Mental State Examination [11]
• The Graded Naming Test [12]
• Wechsler Memory Scale—Revised [13]
• Rey–Osterrieth Complex Figure [14]
• The Visual Object and Space Perception [15]
• The Pyramids and Palm Trees Test [16]

In the Indian context, the following tests prove to be valid:

• The Western Aphasia Battery would help to classify the extent and type of Aphasia in these patients but supplementary tests need to be used.

• Neuropsychological tests also help in arriving at a diagnosis.

• Model-based tests e.g., Psycholinguistic Assessments of Language Processing in Aphasia (PALPA) provide information on underlying ‘processing’ deficits.

5.9 Informal assessment

5.9.1 Assessment of cognitive and linguistic functions:

1. Semantic memory:
   • Recognition of familiar faces.
   • Draw or color objects from memory, classify photographs using semantic criteria or match pictures of objects based on semantic similarity.
   • Executive functions, verbal fluency, and behavior: Impaired verbal fluency is frequently associated with executive function deficits such as abstraction (interpretation of proverbs, cognitive estimates, explaining similarities and differences), response inhibition, or motor sequencing (e.g. alternating hand movements).
   • The ability to create a list of common animals (“category fluency”) or words beginning with a nominated letter (“phonological” or “phonemic fluency”) can be used to measure verbal fluency.
   • Progressive Nonfluent Aphasia (PNFA) is indicated by decreased letter fluency.
   • The amount of words produced in one minute is used to determine the score.

2. Spontaneous speech:
   • The patient can be asked to describe a scene in a photograph or drawing, and the system will analyze an extended sample of the patient’s spontaneous (propositional) speech.
   • This is preferable to asking the patient to describe a typical day in their life.
• Observing the patient’s overall demeanor and demeanor during the clinical interview.

• Frontal dementia is a type of dementia that affects people in a passive way.

• PNFA is a group of people who are frustrated by their issues and try to compensate by using a lot of nonverbal gestures.

• Semantic dementia is characterized by a constant stream of circumlocutory speech.

3. Naming:

• Naming should be assessed both in response to pictorial items (confrontational naming) and in response to verbal descriptions (e.g., “a large grey animal with a trunk”).

• The performance of naming should be evaluated for both high and low-frequency words (e.g., “shoe” versus “moat”).

• Examine if phonological (initial letter) or semantic (related item) cueing improves performance.

• On confrontational naming tasks, visual perception deficits emerge as “visual” errors.

• Circumlocutory reactions, semantic and phonological paraphasias can be noted.

• Cueing with the first letter of the target word may help people with primary word retrieval and phonological encoding impairments (rather than primary verbal store faults).

• Personal names might be particularly challenging.

4. Speech comprehension:

• Word-finding and language output difficulties coexist.

• Can be measured at the level of individual words, which is dependent on both intact perceptual systems and the verbal knowledge store (vocabulary), and sentences, which is dependent on the ability to maintain verbal information online and understand grammatical relationships between words.

5. Single-word comprehension:

• Can be proven at the bedside by testing phoneme pair discrimination (for example, “pat—tap,” and “gat—cat”).

• Noun comprehension—asking the patient to point to items mentioned or described by the examiner, to provide a definition or other information regarding a target word (e.g., “What is a squirrel?”), or to choose between possible synonyms for a target word (e.g., does “trench” mean “hedge” or “ditch?”).
• Asking the patient to classify items according to predetermined criteria (for example, “Is a lion a mammal?”) can be used to dig further.

• When more fine-grained classifications are impossible, meaning is usually kept for broad categories of nouns.

• Comprehension of verbs can also be tested by having the patient choose an appropriate description of the examiner’s actions (“pushing” versus “pulling”, “catching” versus “throwing,” etc.) or by asking them to do acts suggested by the examiner.

• Gestures can also be utilized as an assessment technique.

6. Sentence comprehension:

• A short sequence of acts following unique grammatical norms (e.g., “put the paper beneath the pen that is on the book,” “you take up the watch, then give me the book”) could be used to examine this.

• The patient could also be asked to recognize an image based on a syntactical sentence description (for example, “point to the boy being chased by the dog”).

• Grammar comprehension can be separated into two types: syntactical (word relationships) and morphological (word changes in response to grammatical context), each with its own neurological foundations.

• You can test this by having the patient look for grammatical errors in written sentences.

• An early selective deficit in comprehending grammatical relations may be found in PNFA whereas in SD, comprehension of syntactical constructions is typically intact within the limitations of reduced vocabulary.

7. Speech repetition:

• Patients with impaired speech processing (such as word deafness) as well as those with impaired speech output are at risk.

• It’s possible to assess it at the word and sentence level.

• Single-word repetition is frequently preserved in SD, although sentence repetition is influenced by understanding level.

• When individual words are lost, phonemes may “migrate” between words (for example, “the flag was colored bright red” may become “the blag was fullered with a right breg”), implying that the utterance is encoded as an extended sequence of phonemes (and thus susceptible to re-ordering), rather than a series of meaningful units.

8. Reading, writing, and spelling:

• The patient should be asked to read a piece aloud that contains both irregular and non-words (e.g. proper nouns).
• When reading a passage aloud, the sorts of errors produced convey information about the underlying reading fault.

• Rather than a core language impairment, patients who demonstrate letter-by-letter reading have a problem processing visual word forms: a syndrome of higher-order visual perception (the input to the verbal lexicon).

• Patients with verbal knowledge store deficits (in particular, SD) frequently “regularize” irregular words (e.g., reading “yacht” as “yatched”): this is a “surface dyslexia” [15, 17], in which reading is based on superficial rules for translating written words to speech sounds, rather than a learned vocabulary that governs the pronunciation.

• Regularization issues are particularly obvious for lower-frequency phrases.

• The loss of spelling vocabulary is a defining feature of the SD condition. Despite appropriate noun rendering, phonological dysgraphia (impaired spelling by sound) produces difficulties writing grammatical function words and non-words in PNFA [18].

• Written expression is often better retained with fewer errors than speech in patients with primary speech production impairments (for example, early in the course of PNFA).

9. Sentence generation and completion:

• Once other linguistic functions have been established, this level can be judged with confidence.

• Tasks that demand the development of novel verbal thought, such as the production of a statement including a target word (e.g. “boat”) or the completion of an unfinished phrase, can be used to test the deficiency.

10. Motor assessment:

• It is helpful to distinguish these from any linguistic loss, as well as to progress the clinical diagnosis in general.

• The patient can be instructed to repeat a single syllable quickly (for example, “pa, pa, pa...”) [19, 20]

• Dysarthric patients’ performance will be wrong if their rate or rhythm varies, although AOS patients’ performance is normally normal.

• AOS has a higher rate of sequencing mistakes.

5.9.2 Assessments designed for differential diagnosis and tracking severity in PPA

• Nine neuropsychological evaluations were created or altered expressly for the diagnosis or characterization of PPA, according to a recent systematic review. Several of these can be used to distinguish between clinical variations, and two of them were created expressly to assess PPA severity and progression.

• The Sydney Language Battery (SydBat) is a short battery of tests (image naming, word comprehension, semantic association, and repetition) that can be used to distinguish between PPA subtypes (80% accuracy).
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• The Repeat and Point Test requires patients to repeat 10 multisyllabic words and point to the goal among semantic and phonological distractors in order to distinguish between semantic and nonfluent versions (100% accuracy).

• The Progressive Aphasia Severity Scale (PASS) is a tool used to assess symptoms and follow progression in people with PPA. On a three-point scale, clinicians rate the severity of speech and language deficits (articulation, fluency, syntax/grammar, word retrieval/expression, repetition, auditory comprehension phrases/sentences, single-word comprehension, reading, writing, and functional communication) as well as pragmatic aspects of communication. After an informant fills out a questionnaire and a structured interview with both the patient and the informant, the SLP completes the scale. Table 1 describes PASS.

• The Progressive Aphasia Language Scale (PALS) includes clinician ratings of speech-language features (motor speech and grammatical features in spontaneous speech, naming, single word repetition and comprehension, and sentence repetition and comprehension), but it is based on signs observed during a prescribed set of speech-language tasks rather than symptoms reported via interview or questionnaire. PPA participants were subtyped by variation using an algorithm based on four essential aspects from this evaluation (motor speech impairment, grammar, single-word comprehension, and sentence repetition) (relative to expert clinical diagnosis).

• Clinical Dementia Rating (CDR), a dementia severity rating scale based on a semi-structured interview and clinical judgment, now includes a language domain, which improves sensitivity (relative to the original CDR) for detecting and tracking symptoms and functional impairments in language-prominent dementias like PPA.

5.9.3 Quality of life assessment

1. Communication Activities of Daily Living—3rd edition (CADL-3)

2. Assessment for Living with Aphasia (ALA)

3. Quality of Communication Life Scale (ASHA QCL)


5.10 Probable intervention strategies

5.10.1 Care, support, and treatment team of PPA

1. Neurologist (specialists in brain disorders who make the diagnosis and monitors its symptoms)

2. Neuropsychologist (evaluate cognitive abilities through specialized paper and pencil tests).

3. Psychiatrist (assist with behavioral and mood symptoms).

4. Social worker (help families navigate the difficult decisions related to their diagnosis).
<table>
<thead>
<tr>
<th>PASS Domain</th>
<th>0 = Normal</th>
<th>0.5 = Questionable or very mild impairment</th>
<th>1 = Mild impairment</th>
<th>2 = Moderate impairment</th>
<th>3 = Severe impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>FLUENCY: Degree to which speech flows easily, or it is interrupted by hesitations, pauses, fillers; reduced fluency is accompanied by decreased phrase length and words per minute.</td>
<td>Normal flow of speech</td>
<td>Speech contains occasional blank pauses or use fillers; reduced WPM and/or phrase length</td>
<td>Speech is in short phrases interrupted by pauses or groping for words but it is occasional</td>
<td>Dysfluencies in most utterances; phrase length rarely exceeds three words.</td>
<td>Severely dysfluent speech, phrase length rarely exceeds one word. May not speak.</td>
</tr>
<tr>
<td>SYNTAX AND GRAMMAR: Use of word forms, function words, and word order when forming phrases and sentences in most used modality (speech or writing)</td>
<td>No difficulty in the use of grammar and syntax</td>
<td>Occasional agrammatism or pragmatism (i.e., odd sentence structure such as &quot;I my car drive in your house&quot;) may complain it is effortful to combine words into phrases or sentences</td>
<td>Frequent agrammatism; sentence structures are simple; frequent misuse or omission of grammatical words or sentences</td>
<td>Utterances contain mostly content words with rare use of syntactic words; grouping; functional words or morphological markers</td>
<td>Single word utterances or no speech or writing.</td>
</tr>
<tr>
<td>SINGLE WORD COMPREHENSION: Ability to understand spoken or written a single word.</td>
<td>No difficulty in an understanding a single word in conversation or text.</td>
<td>Occasional difficulty in understanding, low-frequency words (e.g., cork); may question the meaning of words (what?...)</td>
<td>Display lack of word comprehension several times in a brief conversation but able to carry out an easy reasonable meaningful conversation.</td>
<td>Understands some high frequency and / familiar words. Questions the meaning of many words in conversation.</td>
<td>Minimal comprehension of single word comprehension.</td>
</tr>
</tbody>
</table>

Table 1. Three representative domains of the progressive aphasia severity scale (PASS) [21].
5. Speech and language pathologist (provide strategies to maximize communication abilities).


The goal is effective communication, not perfection as PPA is a progressive disorder. Early treatment, a specific plan, partner training, and long-term planning are all critical aspects of proper speech therapy treatment. Speech-language pathologists are uniquely competent to assess the present impact on communication, establish patient priorities, and build a treatment plan based on compensatory methods with a home program to preserve abilities as long as possible. We cannot stop PPA from progressing, but speech-language pathologists can help clients and families adjust their lives, compensate for their impairments, and keep the highest function and quality of life possible.

5.10.2 Tips for treating primary progressive aphasia

5.10.2.1 Practice priority vocabulary

- Practice only what is important to your client.
- Carryover does not occur.
- Create opportunities for your client to practice important words and phrases.
- Use of strategies like “cue cards”.

5.10.2.2 Teach scripts for primary progressive aphasia

- If the client’s main progressive aphasia is still in its early stages, have the family and the client pay attention to everyday routines and words. Request that a family member jots down your ideas. You’ll now have a list of what’s significant in the client’s daily life, and you can use these phrases as scripts to practice as the PPA progresses.
- Speaking scripts can be used for a variety of situations, including:
  - A typical discussions.
  - Making a restaurant reservation.
  - Calls that are made on a regular basis.
- Written scripts/models can be used for the following:
  - Email
  - Texts
  - Check-writing
  - Guidelines for writing effective scripts:

Depending on the severity of the PPA, vary the number and complexity of the scripts—the fewer and simpler the scripts, the more severe the PPA. Concentrate on
clarity and reduce frustration. When in doubt, keep the script simple—you are not trying to teach something new; instead, you are bolstering and compensating. Set priorities based on the client's priorities.

5.10.2.3 Train compensatory strategies for primary progressive aphasia

Identifying and applying communication strengths can help you decide successful compensating measures and increase confidence, just as it can with aphasia or dementia. Modify hobbies and activities so that the person can compensate for linguistic difficulties. Labeling items in the environment is sometimes all that is required. Clients should practice employing techniques, alternative communication, and scripts with the SLP, their family, and close friends in comfortable contexts. It will be easier to use them in public as a result of this.

5.10.2.4 Create a PPA business card

Individualized information cards may be beneficial to clients who are still self-sufficient in the community. This card can be used to inform people that the individual has aphasia, as well as what tactics can help and who to contact in an emergency. This can come in handy if the individual is having trouble at the grocery store or library, or if they have a run-in with the cops. Cards can also be used as a script for ordering a cup of coffee or checking in for tea time.

- To make contacts with new communication partners easier, hand out a customized card that describes what PPA is.

- Make sure the card is in a familiar position for the client, such as their pocketbook or purse, adjacent to their driver's license or photo ID. The card should include any relevant information that the person feels comfortable revealing, such as:

  ○ Name
  ○ Explanation of the communication difficulty or diagnosis (consider including that they are not drunk or on drugs, as is often assumed)
  ○ Quick communication tip strategy that can help (e.g. “use short phrases”, “speak slowly”, “allow extra time to understand”, “be patient”).
  ○ Emergency contact person's name and phone number.

5.10.2.5 Train nonverbal communication

Nonverbal skills become even more critical as PPA advances and communication abilities (especially verbal skills) deteriorate. Nonverbal communication is frequently easier to comprehend and produce. Teach care partners how to use gestures, eye contact, and body language and how to pay attention to them. Allow for more thinking time, have them write keywords, use visuals (photos, drawings, objects), shorten their phrases, and so on.

5.10.2.6 Voice banking and AAC

When words fail, care partners can use AAC apps to help communicate their message. Early on in therapy, you can try out different AAC apps to discover which
From the first day of treatment, the focus should be on a home program and compensating methods. Set up routines and experiment with multimodal and alternative communication as soon as possible, even before the client requires it. It will be easier to understand and acquire new strategies and routines early in the disease process, while more language and cognitive skills are still intact. Clients and families have more time to become comfortable with communication options and select what they like and do not like with earlier involvement. If at all possible, follow up with the client after discharge or check in with them every few months. PPA’s home program and communication tactics will need to evolve as his or her life progresses.

5.10.3 Phased treatment

Clinicians need to consider important differences when managing speech and language impairments in the context of progressive disease [22].

In aphasia caused by stroke, clients show a static or gradually improving communication profile. But clients with PPA will eventually become mute or speak largely in jargon—using words that have some meaning to them but not to the listener.

Ultimately, with the disease’s progression, patients will show increasing cognitive, behavioral, and motor deterioration.

Given this trajectory, PPA treatment should address a client’s current profile of spared and impaired abilities, while also addressing the client’s inevitable decline in communication and, in later stages, other domains.

Because of the evolving nature of PPA, a phased treatment approach, with recurring assessment and repeated treatment “doses” tailored to the client’s current communication needs and challenges is recommended.

5.10.3.1 Phase 1—restitutive treatment in mild PPA

• A crucial part of the SLP’s role during the early stages of PPA is to thoroughly evaluate and document the patient’s communication. This initial evaluation serves as the baseline for the progression of difficulties and helps to identify targets for treatment.

• Clinician often focuses on rebuilding impaired communicative abilities via recitative treatments.

• Treatment during mild stages is critical for helping clients relearn lost skills.

5.10.3.2 Phase 2—toward aided communication in moderate PPA

• Treatment shifts toward multimodal communication and the use of augmentative and alternative communication (AAC).

• Select functional phrases and words to include.

• Strategies to maximize conversational success.

• Also provides counseling regarding PPA.
5.10.3.3 Phase 3—environmental support and communication partner training in moderate-severe to severe PPA

- Treatment focuses on communication partner training and environmental modifications.

- This phased treatment approach allows clinicians to draw from their experience treating aphasia while taking into account the inevitable decline associated with PPA.

5.10.4 Promoting self-cueing strategies

SLPs should focus on self-cueing strategies for lexical retrieval in the mild phases of PPA, which take advantage of the individual’s residual skills [4]. During the examination, semantic, orthographic, and phonological self-cueing should all be tested in order to discover which method is most effective in prompting retrieval.

Family members can be taught to use cueing tactics like “Tell me about it” or “Can you think of what letter it starts with?” to help initiate self-cueing. The ultimate goal is for the person to be able to self-cue at the conversational level on their own. The SLP must be aware, however, when the family member is working too hard or seeing all interactions in therapy sessions.

5.10.5 Script training

Researchers have shown that conversational scripts can help people with stroke-induced aphasia improving their functional communication [23, 24]. The advantage of script training is the automaticity with which patients acquire skills by recalling memories of context-bound, skilled performance. When the same stimuli from the practice environment are available in a functional context, repeated practice of a specific task might improve automatic retrieval. The key to script training is to make sure that each person practices consistently and thoroughly. The SLP should consider the individual’s requirements and interests, the style of the script (dialog or monolog), the number and duration of conversational turns, as well as the grammatical complexity and vocabulary choices when writing a script [25]. Families may purchase a script computer program such as Aphasia Scripts.

Examples of possible script topics include:

- Talking to a family member or friend (e.g. by phone).

- Scheduling an appointment; asking for directions.

- Stating personal information or explaining PPA.

- Giving a lecture or speech

- Saying prayers.

SLPs can frame a written goals based on the accuracy and rate of production (e.g., timing how long it takes to read the script while counting the number of errors). If the person is not motivated to practice the script outside of therapy sessions, he or she may not be a good candidate for script training.
Facilitating receptive language

Facilitating the use of Receptive Language Family training on ways to aid comprehension is critical for individuals with receptive language difficulties. Automatically speaking communication partners can make a difference in comprehension.

• Talk softly, face the person, and give them your undivided attention.
• Augmenting speech with motions.
• Only giving one command at a time.
• Employing grammatical structures and terminology that are easy to understand, or.
• Removing distractions from crucial conversations (e.g., turning off the TV/radio).

Low-tech AAC

The SLP’s job is to determine the book’s proper format, font size, and word/page length, and then instruct the individual and family members to keep adding pages.

• When creating a communication book, SLPs should think about a few things.
• SLPs can laminate small photographs so that the patient can attach them to a key ring and carry them in his or her pocket.
• SLPs should utilize plastic page protectors in a binder to create a more thorough book.
• Using images from the Internet or the person’s own photographs, SLPs can personalize pages.
• SLPs can break the book into sections using tabs.
• SLPs should ask family members to record words and ideas that have led to communication breakdowns. They should add these to the book.

Because some individuals with PPA may not independently use the book, training the family members on appropriate cueing strategies to initiate book use and facilitate navigation also should be targeted during treatment sessions.

High-tech AAC devices

Despite the lack of evidence on AAC intervention options for people with PPA, SLPs have effectively used high-tech AAC devices to help people with severe aphasia caused by stroke communicate [26]. Because people with PPA’s other cognitive abilities deteriorate with time, it’s crucial to keep in mind that a person’s capacity to learn to utilize high-tech equipment may be limited and eventually hampered. The SLP’s job is to figure out which devices if any, will best enable the client to convey
his or her needs and allow for social communication. The SLP should ask a series of questions to determine if a high-tech AAC is a good fit for someone with PPA.

• Is the person enthusiastic about using a high-tech ACC to communicate? Otherwise, it is unlikely to succeed.

• Can the person operate other types of technology independently, such as a computer, microwave, TV remote, or phone? If not, this could suggest a loss of fine motor skills or an inability to sequence, indicating that you are not a good candidate for an ACC.

• Has the family member(s) been involved in the planning, training, and achievement of the objectives? If not, invite family members to several sessions to ensure that the plan is implemented successfully.

• Has the person and their family been taught how to utilize the device in everyday situations? Have the trials attained their objectives? If not, consider additional training and a long trial.

• In conclusion, people with PPA have a variety of choices for increasing communication. Patients and their families are advised to follow the following guidelines:

  • In the early stages of the disease, a comprehensive evaluation of speech, language, and other cognitive abilities should be sought.

  • Follow-up evaluations are required on a regular basis to evaluate patterns of language deterioration.

  • Treatment should begin as soon as possible, focusing on the areas of speech and language that are affected.

  • As language abilities deteriorate, the treatment focus should be modified.

  • Early on, augmentative communication strategies should be implemented. Patients who are given these tactics later in their language loss may find it difficult to learn how to use them.

  • Involving family members or other people with whom the patient talks are critical not just for increasing awareness of effective communication tactics, but also for practicing them with the patient.

  • As patients’ ability to communicate fades, they will rely increasingly on augmentative communication tools. With individual patients, some techniques may be more effective than others, and some patients may utilize more than one.

  • Treatment will not stop the aphasia from progressing, but it will considerably improve communication abilities.

6. General suggestions for communicating with individuals with PPA

*Communication is usually most successful in a peaceful, calm, relaxed situation. Avoid talks when the television is on, and avoid circumstances where
numerous individuals are talking at the same time. Avoiding social circumstances is not necessary; rather, such activities should be encouraged. Maintain usual activities as much as feasible. When people are not tired, communication is usually the most effective.

<table>
<thead>
<tr>
<th>Communication partner</th>
<th>Individual with PPA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Be patient—give your partner time to communicate.</td>
<td>Remember, your partner is trying, so be patient.</td>
</tr>
<tr>
<td>The main goal is the communication of information—not speech or perfection. Once information has been communicated, move on.</td>
<td>The main goal is to communicate—not speech or perfection. Use whatever methods you can to get your message across—speech, writing, drawing, gesture and/or communication notebook. Ask for help when needed.</td>
</tr>
<tr>
<td>Be careful not to speak too quickly.</td>
<td>Take your time.</td>
</tr>
<tr>
<td>Ask questions one at a time, pausing between each. Ask yes/no questions, or multiple choice questions.</td>
<td>Try to give as much relevant information as possible. Focus on expressing the “main points.”</td>
</tr>
<tr>
<td>Repeat what you understand to clarify.</td>
<td>Ask your partner to repeat if you do not understand what was said. Use facial expressions to indicate when you misunderstand.</td>
</tr>
<tr>
<td>Treat the individual as an adult. Do not talk down. Intelligence is generally intact; communication is the problem.</td>
<td></td>
</tr>
</tbody>
</table>

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