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A PRELIMINARY STUDY INVESTIGATING THE NAMING AND NARRATIVE DISCOURSE SKILLS IN THREE VARIANTS OF PRIMARY PROGRESSIVE APHASIA (PPA): SEMANTIC, LOGOPENIC, AND NON-FLUENT/AGRAMMATIC

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Abstract

Introduction: Primary Progressive Aphasia (PPA) is a diagnostic term that refers to the progressive deterioration of speech and language skills while cognitive skills are preserved. The aim of this study was to explore the naming and narrative discourse skills of participants diagnosed with one of the three subtypes of primary progressive aphasia (PPA).

Methods & Procedures: Four participants with a primary diagnosis of primary progressive aphasia participated in this descriptive study. Each participant was administered a battery of cognitive and language measures including the Standardized Mini-Mental State Examination (SMMSE), the Boston Diagnostic Aphasia Examination (BDAE), and the Pyramids and Palm Trees Test (P & PT).Each participant was asked to complete a narrative discourse task using the Cookie Theft picture from the Boston Diagnostic Aphasia Examination (BDAE).

Results: The preliminary results of this study revealed that: the participant with Semantic PPA scored significantly lower on the SMMSE and the P & PT Test. In addition, this participant had a decreased MLU during the narrative task as exemplified by a decreased number of utterances and morphemes. The participant with Logopenic PPA had no impairments in memory or cognition as indicated by their score on the SMMSE. One participant with Agrammatic PPA (A1) did not have any impairment in memory or cognition compared to the other participant with Agrammatic PPA (A2), who had mild impairment in memory and cognition.

Discussion and Conclusions: The results of this study revealed information about the core deficits of the participants diagnosed with the subtypes of PPA.

Keywords: aphasia, primary progressive aphasia, adults, language

Introduction

Primary Progressive Aphasia (PPA) is a diagnostic term that refers to the progressive deterioration of speech and language skills while cognitive functions such as memory, reasoning, and visuo-spatial skills are preserved (Mesulem, 2013). The American Speech-Language and Hearing Association

(ASHA) defines PPA as "A focal dementia (or focal cortical atrophy syndrome) characterized by gradual loss of language function in the context of relatively well-preserved memory, visual processing, and personality until the advanced stages" (American Speech-Language and Hearing Association, 2015). Symptoms of PPA typically begin with word-finding difficulties and later progress to include impaired grammar (syntax) and comprehension skills. A diagnosis of PPA can be determined following a 2-year decline in language functioning which is not accompanied by any marked decline in other cognitive functions. However, it is very important to note that a two year "hiatus" in making a definitive diagnosis should not delay proactive management and treatment of the aphasia (ASHA, 2015).

PPA is caused by a degeneration of the frontotemporal brain region; yet, the person did not suffer from a stroke or CVA. PPA is not easily diagnosed and often involves ruling out clinical syndromes by physicians. The distinct patterns of language deterioration may provide insight into language-specific functions of the brain (Hillis et al., 2006). There is limited research to date about PPA, the different subtypes of PPA, and the language variability and deterioration in the different subtypes of PPA. In addition, there is little known about the discourse and pragmatic abilities of individuals with fluent and non-fluent subtypes of PPA. This study explored the naming and narrative discourse skills of patients diagnosed with PPA.

The three subtypes of PPA include logopenic, semantic, and non-fluent/agrammatic variants. Hillis et al. (2006) note that various types of primary progressive aphasia have been associated with distinct areas of atrophy and pathological changes in the brain. Based on the specific areas of the brain affected, there are differences in the patterns of language deterioration. The logopenic variant of PPA includes core features of word retrieval and sentence repetition deficits. Anomia and single-word comprehension deficits are core features of the semantic variant of PPA. Someone diagnosed with the non-fluent/agrammatic variant PPA typically has agrammatic language skills and effortful speech production. Difficulty recalling the common names of objects is one of the hallmarks of brain damage. Naming impairment observed in degenerative diseases appears to be influenced by other variables including object familiarity (Gaillard et al. 1998).

Previous studies have noted that individuals with PPA appear to progress from one clinical subtype to another, or more commonly, gradually accumulate features of all of the variants (Gorno-Tempini et al., 2004). Such findings support the notion that the variants of PPA are manifestations in the same disease but begin in different areas of the brain (Kertesz et al., 2005). The clinical signs and symptoms provide evidence for the location, more reliably than for the nature, of the brain pathology (Hillis et al., 2006). These previously reported findings, paired with distinct patterns of atrophy, are a basis for speculations regarding the relationship between clinical syndrome, site of neuropathy, and the partial patterns of language deterioration (Hillis et al., 2006).

In her master's degree dissertation, Peacock (1998) observed subjects with PPA and Alzheimer's disease (AD) and noted that the language impairments of PPA are often considered "isolated," whereas the language disturbances of subjects with AD are but one aspect of a pervasive cognitive decline. Sajjadi et al. (2012) recognize the importance of connected speech in the

diagnosis and further classification of PPA. Furthermore, it has been difficult to compare the speech samples of this population because researchers have not adopted a single elicitation method. Therefore, this study attempts to use multiple elicitation methods and probe tasks so that the results may be comparable to other published data findings.

Historically, naming and narrative discourse using pictorial stimuli have been the traditional means of assessment for many researchers working with adults diagnosed with neurodegenerative disorders **(**Bottenberg, Lemme, & Hedberg, 1987; Nicholas & Brookshire, 1993; Petochin, Nicholas, & Brookshire, 1987). Previous research indicates that single pictures are likely to elicit a descriptive discourse genre, in which nouns and verbs are listed with little or no connection between them (Bottenberg et al., 1987). When participants view pictures with actions or complex pictures, they are more likely to produce a narrative rather than simply listing items in the picture. Certain complex pictures that depict a complication or climax may allow the person to infer what happened prior to the climax and what may happen after the climax (Nicholas & Brookshire, 1993). Rogers and Alarcon (1998) recognize a need for further research investigating the decline in speech and language skills in patients diagnosed with PPA. This research is necessary for developing appropriate intervention plans with these patients. There is a critical need for research in this area due to a lack of evidence-based practices (EBP) with this population.

The aim of this study was to explore naming and narrative discourse skills of patients diagnosed with PPA. Discourse is a language unit in which organization supersedes any single-word or sentence. It is the relationship between and among words and sentences, which contributes toward the organization of discourse (Olness, 2006). To date, there have been no published efficacy studies about naming and narrative discourse in persons diagnosed with PPA. These findings may help clinicians and caregivers manage the symptoms associated with PPA by taking a focus on the core deficits of each subtype.

Materials and Method

This study examined four participants with a confirmed diagnosis of PPA. It was approved by the Institutional Review Board at Adelphi University. Case studies are a Level III Evidence Base in Speech-Language Pathology; however, the diagnosis of PPA is difficult to determine and there is limited research with this population to date.

Participants

A total of four participants (n=4) with PPA were included in this study. All participants had a primary diagnosis of primary progressive aphasia from a neurologist. Participants were classified as either semantic, logopenic, or non-fluent/agrammatic. Participants' ages were between 53 and 67 years of age. No participant included in this study had a significant medical history or previous CVA, TBI, or other brain trauma. Participants were recruited via local aphasia groups as well as local hospitals, rehabilitation and skilled nursing facilities.

Participant with Semantic Variant PPA (S1)

S1, a 53-year old female, presented with semantic variant PPA. She was first diagnosed with PPA two years prior to the current study. S1 reported having difficulty with word-finding and often utilized description of objects to communicate when she could not retrieve a target word. S1 did not have a significant medical history and currently receives speech-language intervention from an ASHA certified

speech-language pathologist once a week for thirty minutes.

Participant with Logopenic Variant PPA (L1)

L1, a 61-year old female, presented with logopenic variant PPA. She was first diagnosed with the degenerative disease three years prior to the study. L1 received two years of speech-language therapy intervention prior to testing. However, it was reported that despite medical and therapeutic treatment to remediate the symptoms of PPA, L1's symptoms have gradually worsened.

Participant with Agrammatic Variant PPA (A1)

A1, a 67-year old male, presented with a diagnosis of agrammatic variant PPA. He was accompanied to the evaluation by his wife. He has a significant medical history for high blood pressure, diabetes, and prostate cancer (including radiation). A1 reported that he currently uses an alternative and augmentative communication device (AAC) to express his wants/needs. His device was programmed prior to the evaluation. He independently navigated his device for answers to questions during the study. His expressive language skills consisted of disfluencies. His speech was characterized by syllable and whole word repetitions and fillers such as um, and, uh, and well.

Participant with Agrammatic Variant PPA (A2)

A2, a 66 year-old male presented with a diagnosis of agrammatic variant PPA. He reported that he does not recall when he was first diagnosed. He indicated that he currently receives speech-language intervention. A2 was accompanied to the evaluation by his daughter. Although he interacted and responded to the clinician with socially appropriate pragmatic skills, his responses were sometimes vague and lacked content. A1 stated that he had a significant medical history for diabetes and is currently taking prescription medications to remediate his symptoms.

Measures

In this descriptive study, each participant was administered a battery of cognitive and speech- language measures in the same order.

Standardized Mini-Mental State Examination (SMMSE) Independent Hospital Pricing Authority (IHPA). (2014).

The Mini-Mental State Examination (SMMSE) is a screening method examining cognitive function which takes approximately 10 minutes to administer. The scores reveal information about the possible stage of Alzheimer's disease and the area(s) of impairment. The test includes 11 questions that measure five areas of cognitive functioning including orientation, registration, attention and calculation, recall and language. Each section of the test involves a series of questions or directions for the participant. The participant receives one point for each correct answer (30 maximum points). A score below 23 indicates a cognitive impairment.

Boston Diagnostic Aphasia Examination (BDAE-3) (Good glass, Kaplan, & Barresi, 2000)

The Boston Diagnostic Aphasia Examination (BDAE-3) is a diagnostic tool that determines the type and severity of aphasia. For this study, the principle investigator administered the following sections of the BDAE: (1) Fluency, (2) Conversation/Expository Speech, (3) Auditory Comprehension, (4) Articulation, (5) Paraphasia, (6) Recitation, (7) Repetition, (8) Naming, (9) Reading, and (10) Writing.

The BDAE-3 is used in combination with the Boston Naming Test to detect even mild word retrieval deficits. The BNT is a

component of the BDAE that measures visual confrontation naming.

The Pyramids and Palm Trees Test (P&PT Test) (Howard & Patterson, 1992)

The Pyramids and Palm Trees Test is a test which assesses how a person can access meaning from picture and word stimuli. Information obtained from this test allows the examiner to determine whether a subject's difficulty naming or pointing to a named picture is due to a difficulty in retrieving semantic information from pictures, difficulty in retrieving semantic information from words, or in the case of a naming failure, a difficulty in retrieving the appropriate spoken form of the word (Howard & Patterson, 1992). Six different versions of the test are possible by using a combination of pictures and written or spoken words to change the modality of stimulus or response items.

Procedures

All the participants were seen individually in a quiet therapy room and the test battery was administered in the same order for each participant. Each participant completed an intake form which asked questions about the participant's previous medical history, current diagnosis, and services they were receiving. Afterwards, each participant was administered the testing protocol in the same order: (1) Mini Mental Status Examination, (2) Boston Naming Test, (3) The Pyramids & Palm Trees Test, (4) Boston Diagnostic Aphasia Examination, and the (5) Cookie Theft Picture Description Task.

After the cognitive and language testing was completed, the participant's narrative discourse skills were assessed. The participant was shown the "Cookie Theft" picture from the BDAE. The participants were instructed to look at the picture and then tell the investigator a story about it. The participants were encouraged to use complete sentences and asked one time if they could tell the investigators anything else about the picture scene. The participant's narratives were video-recorded and transcribed in English orthography by the first author of this article. Transcribed narratives were analyzed using a Quantitative Production Analysis (QPA) (Saffran, Berndt, & Schwartz, 1989). The QPA was used to analyze sentence production. It yielded reliable results across raters and across samples for the same participant (Saffran, Berndt, & Schwartz, 1989). The narrative samples were analyzed for the Mean Length of utterance (MLU), total number of utterances/total number of utterances included in the transcription, total number of words/total number of words included in the transcription, total number of morphemes, and number of significant pauses.

Results

Participant with Semantic Variant PPA (S1)

S1 received a score of 18 out of a possible 30 on the SMMSE. This score indicates that she demonstrates deficits that are indicative of cognitive impairment. On the P&PT test of semantic access, S1 scored a 41 out of a possible 52. Scores in this range reflects a deficit in semantic access to words via picture stimuli. S1 obtained a score of 43 on the BNT which falls -3.05 standard deviations below the mean. In her picture description, S1 produced only 7 utterances, 5 of which were included in the final transcription. Her MLU was 4.7 for the included utterances. A total of 26 words and 33 morphemes were included in the final transcription. S1's narrative lacked cohesion as she stated, "the children are helping him, him, her I mean." She demonstrated word-finding difficulties as well as self-corrections throughout the task. In addition, she repeated parts of her oral narrative several times before completing the task.

Participant with Logopenic Variant PPA (L1)

L1 received a score of 26 out of a possible 30 on the SMMSE. Her errors on this measure included deficits in orientation to place and memory for linguistic information (sentence recall). On the P&PT test of semantic access, L1 scored 49 out of a possible 52. Her score was within the normal range and did not indicate deficits for semantic access to picture information or words. The BNT and BDAE were not administered to L1 because she had recently undergone a testing battery which included these test measures. In addition, L1 appeared to become fatigued during testing and the investigator discontinued the session. L1's narrative was short and lacked details. In addition, she used circumlocutionary behaviors during the production of her narrative. When looking at the picture stimuli, she described the objects in the picture but did not provide the investigator with a sequence of events in the story. Her narrative primarily consisted of nouns as she named different people/objects. However, L1 demonstrated the highest MLU at 8.6 compared to any other participant. She did not have significant or increased amount of pause times. During her production of six utterances, L1 produced 63 words and 78 morphemes.

Participant with Agrammatic Variant PPA (A1)

A1 received a score of 29 out of a possible 30. His error occurred on the sentence repetition task. It was determined by the examiner that his disfluencies prevented him from completing this task rather than an actual deficit in the area of memory for linguistic information. On the P&PT test, he received a score of 34 out of a possible 52. A1 had significant difficulty with this task. Given extended response time, he continued to struggle to make semantic associations between the picture stimuli. On the BNT, S1 scored a 55 which is equivalent to +.37 standard deviations above the mean. It is important to note that for this task, A1 was provided with multiple choice written responses to choose from since he could not use his device for this task. The written responses may have aided him in recalling the names of different objects. In his picture description, A1 produced 22 utterances, 10 of which were included in the final transcription. Many utterances were omitted according to Brookshire's rules of transcription and QPA coding. His narrative included 62 words and 75 morphemes. He demonstrated an MLU of 7.5 and did not have significant pause times between words or utterances.

Participant with Agrammatic Variant PPA (A2)

On the SMMSE, A2 received a score of 50 out of a possible 52. His errors included sentence repetition and following written commands. Although he read the written command correctly, he did not perform the action (i.e. close your eyes). On the P&PT test of semantic access, A2 scored a 50 out of a total 52 responses. He did not demonstrate significant difficulty with this task. On the BNT, he received a score of 49 which is equivalent to -.93 standard deviations below the mean. Although he scored below the mean on this test measure, his score was still within normal limits for his age. During the picture description task, A2 produced a total of 16 utterances. Ten of the 16 utterances were included in the final transcription. He produced 63 words and 71 morphemes. Although he related content information from the event, his narrative did not follow conventional norms. He often paused to ask questions and reiterate something humorous about the picture scene. He did not create a story about the events being depicted in the scene.

Table No. 1: Participant Characteristics and Test Scores

PPA Subtype	Age	Gender	SMMSE Score	P&PT Score	BNT Score
Semantic (S1)	53	Female	18/30	41/50	43 (-3.05 SD)
Logopenic (L1)	61	Female	26/30	49/50	Not Tested
Agrammatic- (A1)	67	Male	29/30	34/50	55 (+.37 SD)
Agrammatic- (A2)	66	Male	24/30	50/50	49 (-.93 SD)

Table No. 2: QPA Results for all the Participants

	S1	L1	A1	A2
Total # Utterances	7	9	22	16
Total # Utterances Included	5	9	10	10
Total # Words Included	26	63	62	63
Total # Morphemes	33	78	75	71
# of Significant Pauses (> 2-3 seconds)	1	0	0	2
Mean Length Utterances (MLU)	4.7	8.6	7.5	7.1

Discussion and Conclusion

The results of this study revealed some insight into the deficits associated with each subtype of PPA. The participant with semantic variant PPA scored significantly lower on the SMMSE and the P & PT Test compared to the other participants diagnosed with the other subtypes. The investigators posit that this occurred because a core deficit of semantic variant PPA includes difficulty following directions. In addition, these participants typically have difficulty in naming or pointing to a picture which may be due to a difficulty in retrieving semantic information (meaning) from pictures. These participants also have difficulty producing a complex narrative. The narrative produced by this participant had a decreased MLU as exemplified by a decreased number of utterances and morphemes.

The participant with logopenic variant PPA exemplified a good SMMSE score. Although the hallmark of this subtype is decreased word-finding skills, this participant did not have difficulty naming or pointing to a picture which indicates that the participant did not have difficulty in retrieving semantic information from

pictures. The participant produced a longer narrative compared to the participant with semantic PPA; however, not as long in comparison to the agrammatic participants' narratives. The narrative produced consisted of more complex structures, including morphemes and content words than the other subtypes.

The two participants presenting with agrammatic variant PPA exemplified good to mild deficits with the SMMSE. They did not have difficulty retrieving semantic information from a picture as indicated by the P & PT Test. The MLU was greater as exemplified by the total number of utterances; however, the total number of included utterances was reduced. There were a large number of morphemes for these participants' narratives; however, the type of morphemes was not analyzed.

Moreover, in analyzing the test results of the four participants included in this study, we were able to learn more about the core deficits associated with each subtype of PPA. By identifying the core deficits associated with each participant's subtype of PPA, we were able to make conclusions that may be transferred to therapy interventions for these participants.

The results of testing revealed several trends associated with each subtype of PPA. These findings may prove valuable for clinicians who are planning their intervention and goals for subjects with a diagnosis of PPA. Similar to Gorno-Tempini et al.'s findings (2004), we observed that it is not uncommon for individuals with PPA to present with a combination of symptoms that are characteristic of various subtypes of PPA. We noted this in the participants who had presented with a PPA diagnosis for greater than 3-5 years. These findings also support the research done by Kertesz and colleagues (2005) who proposed that the variants of PPA are manifestations of the same disease but begin in different areas of the brain. Since the disease has a degenerative course, clinicians must consider the core deficits of each subtype before selecting appropriate goals. Testing methods included in this study proved to be useful assessment tools in identifying a subject's speech and language difficulties.

Limitations and Future Research

The current study had a small sample size; therefore, the results could not be generalized to the larger population of participants diagnosed with PPA. Future research is warranted to investigate the different variants of PPA with a larger sample size. In addition, studies should be designed that examine the long-term effects of PPA on the speech-language and cognitive skills of participants with each variant of PPA. This will lead to better evidence-based intervention approaches for working with this population.

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