

Critical Review:
**The impact of intervention on the maintenance of speech and language skills
for adults with Primary Progressive Aphasia (PPA)**

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This critical review examined the impact of intervention on the maintenance of speech and language skills in adults with Primary Progressive Aphasia (PPA) a neurodegenerative disease characterized by loss of language faculties over time with an onset between 40 to 75 years of age. A literature search was completed and the following types of studies were reviewed: two case studies and five single-subject experimental studies. Overall, the research provided preliminary evidence that a variety of speech and language interventions can help maintain speech and language skills in individuals with PPA. However, it was unclear from this evidence which types of interventions provided the most effective maintenance of speech and language skills due to limitations within the reviewed literature. Therefore, continuing research on effective speech and language interventions for individuals with PPA is warranted.

Introduction

Primary Progressive Aphasia (PPA) is a rare neurodegenerative disease characterized by at least two years of isolated and gradual language decline with initial preservation of other cognitive functions such as memory, visual processing, behavioural function and activities of daily living (Mesulam, 2001). Similar to stroke-induced or 'static' aphasia, PPA has different subtypes including: a) *non-fluent* characterized by telegraphic and/or disprosodic speech containing frequent phonological errors; b) *fluent* characterized by difficulties in the semantic aspects of language with severe anomia but generally intact syntax and fluency; and c) *mixed* both fluent and non-fluent characteristics (Croot & Patterson, 1999, Mesulam & Weintraub, 1992). Typically, PPA affects more males than females (ratio of 2:1) with onset occurring between the ages of 40 to 75 years (Duffy & Peterson, 1992).

The speech and language profiles of individuals with PPA vary widely and these individuals face a poor prognosis. Despite this, many individuals with PPA will strive to remain active and independent following the onset of their language symptoms (Murray, 1998). In order to facilitate this, viable and effective speech and language therapy options need to be provided to these individuals.

Although there are some clear similarities in the speech and language symptoms experienced in both PPA and 'static' aphasia, effective and appropriate methods for the treatment of PPA remain unclear. Minimal research has been completed in this area. In

part, this could be due to the rare and heterogeneous PPA population, which makes group studies difficult as well as the progressive nature of the disease, which makes studying the long-term effects of an intervention challenging. Due to these factors, research within this population may be limited to case studies or single-subject designs.

The typical speech and language interventions for stroke-induced aphasia focus on improvement of skills over time; however, given the progressive decline of skills in PPA, this is not appropriate. Rogers and Alarcon (1998) suggest that therapy should focus on the maintenance or stabilization of existing skills, rather than on the improvement of these skills. In order to accomplish this goal, clinicians need to be informed of the available speech and language interventions and the impact of these interventions on individuals with PPA. The important questions remain: How does one effectively treat an individual with PPA? What will be the impact of a speech and language intervention on this individual?

Objectives

The primary objective of this paper is to critically examine the existing literature to determine the impact of intervention on the maintenance of speech and language skills for adults with PPA. The secondary objective is to provide recommendations for clinical practice.

Methods

Search Strategy

The research articles selected for this critical review were found using a computer database search including: PubMed, Medline, PsycINFO, Scopus and Scholars Portal. The search was not limited to specific years, thus, it included articles published from the earliest possible date to 2008. The following search strings were used: (Primary Progressive Aphasia) AND (Treatment) OR (Speech and Language Treatment) OR (Intervention) OR (Therapy). In addition, hand searches for relevant articles from reference sections were also performed.

Selection Criteria

Studies were included if they focused on speech, language or alternative/augmentative communication interventions for adults (40 years of age and above) with the diagnosis of PPA (time post-onset was not limited). Also, studies that investigated patients with other disorders in addition to PPA were included. Any studies that focused solely on drug or surgical interventions for PPA were not included.

Data Collection

The search strategy identified seven studies that met the selection criteria outlined above. These studies included two case studies and five single-subject experimental studies.

Results

Case Studies

Case studies are often useful as a starting point to direct future research in rare populations such as PPA. However, case studies lack external validity in that the results from single cases cannot be generalized to larger populations.

Rogers and Alarcon (1998) conducted a 4 year longitudinal case study on a 69-year-old man with non-fluent PPA and moderate-severe apraxia of speech (AOS). The main purpose of this article was to document the client's language decline; in addition, the therapy approach used with this client was discussed.

The authors reported a detailed description of the client's personal and medical history as well as results from various speech, language and cognitive assessments completed over the four year period; providing a clear understanding of the client's background. In addition, standardized assessment tools were listed and extra assessment protocols were also described, making replication possible.

Although a significant amount of detail was provided on the assessment protocols and results, little

information was provided on the speech and language therapy used for this client. A 'proactive management approach' to therapy was reported, which involved providing the client with compensatory strategies throughout his speech and language declines. However, no details on the specific therapy activities or the frequency of therapy visits were given in this study; rather, the authors discussed the principles on which this type of therapy was based. No outcome measures linked to the therapy approach were provided. The lack of information on both the therapy approach and outcomes made it impossible to conclude its effectiveness.

Overall, this case study provided anecdotal evidence only on the impact of the therapy approach for this client. However, clinicians may find this article useful for informational purposes when working with similar clients.

One additional issue addressed by this study was the question of how to measure the short-term language declines that occur in clients with PPA. The standardized tests used to measure the decline of speech and language abilities over time were reported to be generally consistent with clinical impressions; however, short-term declines were not accurately reflected in these tests. More sensitive and informal measures of the rate of language decline were derived through the analysis of connected speech. These included, words per minute (WPM), mean length of utterance (MLU) and correct information units (CIUs). The measure of CIU involved a count of intelligible words that were accurate, relevant and informative relative to the eliciting stimulus, although they may not have been produced in a grammatically accurate manner. The authors determined that these measures accurately reflected the short-term language declines observed in the client.

The therapy approach reported in a case study completed by Cress and King (1999) involved the development of multimodality AAC strategies for two individuals with PPA. In the first case, a 59-year-old woman, the authors administered a standard speech and language assessment protocol and subsequently explored options for a communication book. It was reported that this client quickly learned how to use the communication book; however, no outcome measures were provided. In the second case, a 60-year-old man, a standard speech and language assessment was not given as the client refused to participate in the tests. However, the authors based the development of his AAC strategies on information from informal language samples and

designed communication boards for receptive and expressive language.

The authors provided examples of the types of symbols used on both communication boards as well as information on the frequency and duration of the training. This allows for replication of this particular intervention.

In order to measure the outcomes of this client's AAC intervention the authors stated that tape recorded conversations, informal notes and tallies by both the therapist and family members on the use of receptive and expressive symbols were taken; unfortunately, this data was not reported in detail. The authors concluded that this client successfully learned how to use both communication boards, with some situations providing greater success than others (e.g., when used with familiar listeners). It was also stated that the client used more complex messages and took more communicative turns when he used the communication boards.

Overall, the authors stated that there was good potential for using visually based AAC strategies for individuals with PPA based on the increase in functional communication for both of these clients. However, clear outcome measures of both AAC interventions were not provided; therefore, the accuracy of the author's interpretations and conclusions from this study cannot be determined.

Single-Subject Experimental Studies

Similar to case studies, single-subject experimental studies are also useful in studying rare and heterogeneous populations such as PPA. This type of design allows researchers to study the effects of a specific treatment on a client. Also similar to case studies, these types of studies lack external validity; however, the strength of evidence is slightly higher as the client is a part of an experimental study that yields data which can be used to support conclusions and interpretations.

Murray (1998) conducted a 2.5 year longitudinal single-subject experimental design study that focused on an evolving treatment regime for a 64-year-old woman with non-fluent PPA. Within this intervention, three different therapy approaches were used: 1) a traditional stimulation-facilitation approach; 2) the 'back-to-the-drawing-board' program (Helm-Estabrooks & Albert, 1991); and 3) a functional communication approach which included provision of an AAC device.

The author provided specific details on the therapy materials, therapy procedures as well as the

frequency and duration of all therapy approaches making replication possible. In addition, the author reported results from standardized language assessments completed throughout the duration of the study as well as direct outcome measures linked to each therapy approach; a good description of the client's personal and medical history was also included.

An additional strength of this study was in the treatment designs. Specifically, two therapy approaches were implemented in multiple block treatments, which allowed for replication of treatment effects. In comparison to the single block treatment design (used for the 'back-to-the-drawing-board' approach), this design provided additional information on the carryover of skills between treatment blocks. It allowed for the client to be her own control and illustrated the level of her communication skills both with and without treatment, thus clearly indicating the impact of these therapy approaches.

The raw data from both formal and informal pre- and post-treatment measures relevant to each therapy approach were reported, thus increasing confidence in the author's interpretations. Data analyses involved both qualitative and quantitative methods. The quantitative statistical analyses involved pairwise *t*-tests on the pre- and post-treatment measures in only one of the treatment approaches. The remaining quantitative data collected was analyzed using visual inspection by the author. Both types of analyses were considered appropriate given the nature of the data collected.

The author provided results from a qualitative analysis of the client's conversation, but no information was provided on how this analysis was completed, limiting confidence in the accuracy of the author's interpretations of the qualitative data.

The lack of reported reliability values in this study made it difficult to determine if the data was collected in an unbiased manner. This added the limitation of experimenter bias in both the data collection and analysis phases of this study.

Overall, this study provided detailed descriptions of three therapy approaches used for an individual with PPA. Although there were some limitations to this study, it provided a fair degree of evidence in support of the benefits of long term speech-language pathology services for individuals with PPA.

McNeil, Small, Masterson and Fossett (1995) conducted a single-subject experimental study to evaluate the effects of behavioural (cueing hierarchy) and pharmacological (dextroamphetamine) treatments on the lexical-semantic performance of a 61-year-old male with PPA. Overall, they determined that the client benefitted from both the behavioural + pharmacological and the behavioural only treatments. This indicated that it was possible to improve the lexical retrieval abilities of this client, despite an overall decline in other language and communicative behaviours.

The authors reported detailed information on the client's personal and medical history, which provided necessary background information on this client. In addition, extremely detailed descriptions of the treatment design, treatment stimuli and therapy procedures were provided which allowed for replication.

Specific information was given regarding the derivation of treatment word lists, the cueing hierarchy, frequency and length of baseline sessions, duration of each type of treatment, and withdrawal periods. Therapy procedures were also discussed in detail and descriptive information (word frequency and word length) for all of the treated and probe lists were also reported.

The treatment design included multiple baselines, multiple treatment (behavioural, behavioural + pharmacological), multiple probes as well as withdrawal and maintenance periods. This type of design was necessary to accurately determine the differential effects of the behavioural and pharmacological treatment approaches as well as to determine treatment, generalization and maintenance effects. Informal measures directly linked to therapy tasks as well as standardized measures were administered; analyses of connected speech samples (e.g., CIUs) were conducted. These measurements provided a well-rounded evaluation of the client's overall language declines throughout the study.

Two uninvolved judges visually inspected the data and were asked to report whether a treatment, generalization, or maintenance effect had occurred for all informal measures; only the data that the judges agreed upon were included in study. These judges eliminated the potential for experimenter bias in the data analysis, which can often be a limitation in experimental studies.

The main limitations to this study were small sample size, lack of reported reliability and lack of statistical

analysis. The limitation of weak external validity due to small sample size was acceptable given the population of study; however, the lack of reported reliability measures was not. The authors report that the judges were in agreement during the data analysis phase; however, without reported reliability values, the strength of this agreement could not be determined. Also, within the data collection phase, no reliability measures were reported; therefore, it could not be concluded with confidence that the data was collected in an unbiased manner. The authors could have controlled for reliability in the data collection phases by videotaping sessions and having an uninvolved experimenter rate the client's responses.

The lack of statistical analysis was considered a limitation within this study as it was not clear, given the nature of the data, why statistical analyses were not performed.

Overall, given the strengths in the design of this study, it can be concluded that it provides a fair to good level of evidence in support of the improvement of lexical retrieval abilities in this client.

Schneider, Thompson, & Luring (1996) conducted a single-subject multiple baseline experimental design on a 62-year-old female with non-fluent PPA. The purpose of the study was to examine the effects of a verbal plus gestural matrix treatment procedure on the acquisition and generalization of present, past and future verb tenses in simple sentence production. A reversal design was incorporated such that gestural responses were withdrawn after three treatment sessions. Results showed that correct sentence production was more likely when verbal and gestural responses were paired; however, correct verbal responses declined only in the past tense condition after the removal of gestural responses.

Detailed information on the client's personal and medical history was provided which gave a clear overview of the client. In addition, results of pre-treatment standardized speech, language and cognitive assessments were reported, which clearly illustrated the client's deficits.

Specific procedural information was reported regarding all aspects of the study. In addition, the appendices included all of the sentence stimuli, the scoring protocol as well as the verbal plus gestural treatment sequence verbatim making replication possible.

This study involved a multiple baseline treatment design; however, only two sessions were devoted to

establishing these baselines. Out of the three verb tense conditions (past, present, future) a baseline of verbal performance was only established for one. The lack of baselines for all conditions limited the conclusions that could be made about the effectiveness of the treatment.

The authors reported that sessions were videotaped for reliability purposes. Reliability for client responses in baseline and probe sessions was 92%. Inter-judge reliability in training sessions was reported to be 97%. Reporting these values showed that the potential for experimenter bias was eliminated in the data collection phases of the study.

The outcomes directly linked to treatment were reported as the percentage of grammatically correct sentences in both the trained and untrained sentences. In addition, narrative language samples were taken pre-treatment, during and post-treatment to determine the generalization of treatment; results reported from these samples included a variety of linguistic factors (e.g., MLU, mean number of embedded clauses, % grammatical sentences, % simple and conjoined sentences, etc.). These variables were considered to be representative generalization of treatment effects.

The data was analyzed by the authors through visual inspection, which was considered a limitation as no statistical significance was reported. It was unclear as to why a statistical analysis was not completed given the nature of the data.

Overall, this study provided a fair level of evidence to support that the use of gestures facilitates verbal responses. Given the limitations within this study, it was concluded that the authors may have overstated the importance of their findings as verbal responses only declined in one condition after the removal of gestures.

Rogalski and Edmonds (2008) conducted a single-subject experimental design study which targeted the discourse level of a 76-year-old male with PPA. The treatment used in this study was Attentive Reading and Constrained Summarisation (ARCS).

The authors provided a concise and thorough case history of the client including standardized speech, language and neurological assessments; this gave adequate information on the client's background. Specific information was also included on the treatment frequency, stimuli and protocol. The appendices included the treatment protocol verbatim as well as pre- and post-treatment language samples from the client. In addition, the coding procedures

used to determine coherence, cohesion and informativeness/efficiency (i.e., WPM, CIUs) from discourse samples taken at three time points (pre-, immediately post- and two months post-treatment) were provided. Replication was possible based on this reported information.

The discourse samples were reported to be based on the Nicholas and Brookshire (1993) picture description tasks, which provided good session-to-session reliability. The variables analyzed from these samples were considered to be appropriate as they provided information on macro (e.g., topic maintenance) and micro (e.g., word retrieval) structures of discourse, the areas focused on in treatment; however, the data linked directly to treatment were not provided.

Reliability was assessed for all variables and the raters were blinded to the condition (pre- or post-treatment); the inter-rater reliability was good as it was reported to range from 85.7% to 100%. The data was analyzed using visual inspection, which was considered appropriate for this type of data; however, a design allowing for statistical analyses would have strengthened the level of evidence.

Given the limitations, this study provided a fair level of evidence in support of the long-term success of this treatment for individuals with PPA.

Pattee, Von Berg and Ghezzi (2006) conducted a single-subject experimental design study that investigated the effects of two different modes of communication on the communicative output of a 57-year-old woman who was no longer able to communicate verbally due to PPA and AOS. The two treatment approaches were: 1) a text-to-speech alternative communication device (ACD), the Light-Writer; and 2) American Sign Language.

Limited information was provided on the client's personal and medical history as well as the study design and treatment procedures. Incomplete information within this type of study is considered a strong limitation as replication should be possible based on client characteristics as well as the design.

Informal outcome measures directly linked to each communication intervention were reported pre- and post-treatment based on responses to Nicholas and Brookshire (1993) line drawings. These included the number of words/signs used, total CIUs and percent CIUs. These outcome measures were appropriate for the analysis of the discourse sample rendered from the alternative methods of communication.

The data was analyzed through visual inspection by the author; therefore, no statistical significance was reported. In addition, nine graduate students were asked to rate the information content of the client's pre- and post-treatment responses, which eliminated some of the experimenter bias from the study.

The authors included the rating scale that was used for the client to evaluate preference of communication method; however, the raw data from this rating scale was not included. Therefore, the statement that this client responded much better to the ASL communication method based on personal preference was not supported by any data.

Overall, based on the limitations mentioned above, this article provided a fair level of evidence on the use of alternative communication methods for individuals with PPA. This study also raised awareness that personal preferences may play a role in determining which intervention method will be successful for individuals with PPA.

Discussion

All of the studies reviewed in this paper target speech and language therapy on adults with PPA; however, the specific focuses within these studies are very different. All of the studies report some benefit to the client's speech and language skills due to the intervention. However, the strength of evidence from these studies is limited due to a number of methodological limitations. Firstly, there are concerns regarding sample size and participant selection. Each study has only one participant, or the participants are presented as separate cases (e.g., Cress & King, 1999). Also two of the studies (Rogers & Alarcon, 1998; Pattee et al., 2006), involve participants that have both PPA and AOS, rather than a pure case of PPA. These factors limit the generalizability of the findings from these studies to adults with PPA as a group.

Another methodological limitation found within these studies was the lack of statistical analyses. With the exception of the Murray (1998) study where a pairwise *t*-test was reported, no statistical analyses were reported. The visual inspection of data for analysis purposes was appropriate given the nature of the data in some of the studies; however, in others (e.g., McNeil et al., 1995; Schneider et al., 1996) it was not clear why statistical analyses had not been performed. When pre- and post-treatment measures are reported, it is difficult to determine if there is a significant effect of treatment without statistical analyses.

Also, in the case studies (Cress & King, 1999; Rogers & Alarcon, 1998), no specific data were reported, thus the impact or success of treatment was based solely on the authors' interpretations. This makes it difficult to determine the impact of these interventions as the authors may have been biased.

There are also advantages to this type of research. All of the studies, with the exception of one (Pattee et al., 2006) provide detailed descriptions of the clients' case history. This allows for clinicians to use these studies for informational purposes with similar clients.

In addition, the study by Rogers and Alarcon (1998) suggested that standardized tests did not accurately reflect short-term language declines in PPA. They suggested the use of informal measures such as MLU and CIUs from language samples were more appropriate. These informal measures were used in other studies to measure treatment effects (McNeil et al., 1995; Schneider et al., 1996; Rogalski & Edmonds, 2008; Pattee et al., 2006) indicating that this is one effective way to measure language ability in PPA.

Overall, the evidence from this critical review is suggestive. In order to strengthen the level of evidence future research considerations in this area should include larger sample sizes matched for various characteristics (e.g., type of PPA, time since diagnosis, age, gender) and study designs that incorporate statistical analysis.

Clinical Implications

Due to the limited strength of evidence provided from the reviewed articles, clinicians should be cautious when implementing the findings from these studies into practice. However, this review shows that there are many different interventions that can help maintain speech and language skills in individuals with PPA. Clinicians should be aware that treatment within a rare and heterogeneous population such as PPA may require an eclectic approach as effective strategies may vary between individuals. Also, the focus of therapy may have to change over time as the client's skills decline; thus, therapy should be long term to accommodate for these changes. In addition, the success of a speech and language intervention may also depend on the client's personal preferences and motivations.

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