

**Critical Review:  
Effectiveness of Speech Therapy for Individuals with Primary Progressive Aphasia**

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Primary Progressive Aphasia (PPA) is a focal dementia syndrome with deterioration of language functions but relative preservation of other cognitive functions for at least the first two years of the disease. This paper critically reviews nine studies of communication treatments for individuals with PPA. These treatment regimes include behavioural training (phonological and lexical/semantic), training with alternative and augmentative (AAC) techniques, combined therapy approaches, high frequency repetitive Transcranial Magnetic Stimulation (hf-rTMS) and Attentive Reading and Constrained Summarization (ARCS). The outcomes of these studies provide persuasive evidence of the value of speech therapy for individuals with PPA, although more research is required in this area.

### *Introduction*

Primary progressive aphasia (PPA) is a focal dementia characterized by an isolated and gradual dissolution of language (Mesulam, 2001). It is a progressive disorder in which language abilities may decline for many years prior to any manifestation of cognitive symptoms. Expression often is compromised earlier than comprehension, with word finding or motor issues frequently being the first symptoms. Diagnosis of PPA requires that the patient has experienced isolated progressive language decline for at least two years while all other mental faculties, such as memory, visuospatial skills, reasoning and personality remain relatively intact (Mesulam, 2001). Neuropsychological testing is often used for this purpose, but must be compared with clinical findings, as many of these tests have some overt or covert reliance on language.

Brain imaging may also be used in the diagnosis of PPA, looking at atrophy or abnormalities in blood flow and metabolism in the language centres of the brain. (Mesulam, 2001). Language symptoms, especially naming difficulties, may emerge prior to any brain scan abnormalities.

Unlike aphasia post trauma, which can often be matched to a clinicopathological pattern (i.e. Broca's, conduction, etc.), PPA rarely fits into these classical patterns and there is no single type of language dysfunction that is pathognomic for PPA. Word finding difficulty is often the first symptom to appear in patients with PPA. Auditory and reading comprehension is often relatively preserved early on, but generally decays to the point where even single words cannot be understood. Agrammatism, also often present as the disorder progresses, can lead to

the near mutism, or highly telegraphic speech, filled with circumlocutions, fillers, paraphasias and, in the later stages, neologism (Mesulam, 2001). Reading and writing difficulties, while often preserved early on, are compromised in later stages. In the terminal stages of the disease, the production and comprehension of language are both severely compromised, with possible cognitive and behavioural changes at this stage. These highly variable profiles present difficulties with research validity and reliability, making it difficult to collect a base of strong evidence.

This is a topic of clinical and research importance. PPA has a devastating effect on the lives of the patients and their families. It is imperative we use evidence based practice to inform our decisions regarding treatment, both to provide optimal therapy to our patients and to use the limited resources we may have effectively and efficiently.

### *Objectives*

The primary objective of this research paper is to critically review the literature on communication treatments carried out by speech-language pathologists for patients with PPA, examining the effectiveness of speech therapy for this population. The secondary objective of this paper is to provide evidence-based recommendations for continued research in this area, as well as clinical implications for future treatment.

### *Methods*

#### Search Strategy

Internet databases, including SCOPUS: Health Sciences (>5300 journals) and Social Sciences

(>2800 journals) and PubMed (>4500 journals) were searched with the following terms:

(speech language pathology) AND (primary progressive aphasia)  
(speech therapy) AND (primary progressive aphasia)  
(therapy) AND (primary progressive aphasia)

The search was limited to articles written in English. There was no limitation on the date of articles.

### Selection Criteria

Studies selected for review in this critical review were required to address a protocol designed to improve communicative effectiveness, and must include at least one individual diagnosed with PPA. The diagnoses must show isolated language decline without cognitive decline. Outcome measures for the treatment must also be reported for inclusion in the study. No limits were set on the demographics or linguistic profile of the research participants.

### Data Collection

The data search yielded seven papers that fit the aforementioned search criteria. Two additional articles were located using the references of the articles found through the database search. There are eight case studies one hybrid single subject experimental design.

### **Results**

Most of the below studies are single subject designs. With this comes the inherent lack of external validity, as the results cannot be easily generalized to other subjects with the disorder, especially with the varied profiles found in PPA patients. This is an on-going issue in researching small populations, and adversely affects the level of evidence. However, good subject design, coupled with multiple studies can help to increase this level of evidence.

### Behavioural Therapy

Koenig- Bruhin et al (2005) reported a single case study, in which the participant was a 72 year old male, with fluent PPA. He presented with language abilities below normal limits, predominantly expressive, and cognitive abilities within normal limits for his age cohort. The patient was given a 5 month course of therapy (once/week), including daily homework. The therapy focused on phonological therapy combined with short term memory tasks. Pre and post tests were administered, showing a significant improvement on sentence repetition. Naming of infrequent words was improved, but was not statistically significant, although one must take

into consideration the progressive nature of the disorder. No other areas of improvement were considered significant.

These results must be regarded cautiously. The study has little replicability as it provides limited information on the tasks used in therapy, and thus the findings have low reliability. Also, it has limited ecological validity, as sentence repetition, the only area of significant change, has limited use in day to day communication.

Louis et al (2001) reported on three individuals with non-fluent primary progressive aphasia. Patients were comprised of two females and one male, ages 64, 71, and 77. All patients presented with typical non-fluent PPA, involving moderate to severe expression impairments, as well as comprehension impairments as the disorder progressed. The therapy protocol consisted of daily 15-20 minute exercises of tapping syllabic and phonemic segmentation and discrimination skills training. In addition, exercises were recorded on audio and slowed to 166% of the original speech signal, to facilitate the improvement of auditory perception of the rapid transitional elements of speech, in accordance with the temporal theory of phonemic processing. The training period lasted for 43 days, with one participant receiving an additional 30 days of treatment with modified speech exercises. The authors found a gradual and global improvement across the 43 days. Significant improvement in fluency was seen for one participant, written comprehension for one, reading for two, and repetition for two. There was no significant improvement in oral comprehension or naming in any participants. Global decline was seen in other areas (those not presumed to be related to the treated phonological processing) of language in all subjects, as is expected with a progressive disorder. To give further evidence to this hypothesis, participant 1 was given two additional 30 day periods of therapy, the first (period 2) using normal, unmodified speech in the exercises and the second period (period 3) using slowed speech again. A statistically significant difference between these two periods (unmodified and modified speech), was obtained using a repeated measures ANOVA ( $F=2.995$ ,  $p=.0021$ ), with performance in the unmodified condition (period 2) tending to worsen, while it improved in the modified condition (periods 1 and 3).

This study is well designed, and has compelling evidence for the treatment, as the improved results cannot be seen in the condition in which the modification has been removed. Testing of untreated language areas provides a context which the results

can be viewed in against the background of the progressive nature of the disorder, increasing the validity. The results must be viewed warily though, as there may have been other factors associated with the treatment that had an effect on the progression of the disorder, or unknowns about the progression of the disorder itself, which could confound the data. Further study on phonological processing and the effect of modified auditory input are warranted.

Schneider et al (1996) present a single subject multiple baseline study looking at the effectiveness of verbal combined with gestural treatment on the acquisition and generalization of the verb tenses in a subject with PPA using a matrix training procedure. The subject presented with a non-fluent language impairment consistent with agrammatism and showing more severe impairment in spontaneous speech tasks than formal testing. Post-treatment scores showed an improvement in both trained and untrained words and tenses in the context of simple SVO sentences, which were maintained at the 3 months post-treatment, except in spontaneous speech measures. Gestural pairing appeared to have a large effect and was controlled for with a reversal component, in which the subject's scores fell when no gesture was presented.

This is a well designed study, improving its validity as case study. Baseline data, probes, post-treatment and maintenance measures give increased information and improve the evidence base for this treatment. Due to having a single subject, external validity is reduced. It has good construct validity, with use of the NP+V+NP context, as this is a prevalent structure in English, and thus is highly related to the subject's communicative effectiveness. It provides moderate evidence for the treatment of PPA using a matrix design with verb tenses. More persuasive evidence is seen for the pairing of gestures with words, as the reversal component made it clear that gestures had an impact on the subject's ability to use the verb tenses appropriately. The lack of maintenance in spontaneous speech measures must be interpreted cautiously, due to the progressive nature of this disorder. Overall, this study must be regarded with caution until further evidence emerges.

#### AAC Strategies

Cress and King (1999) report on two case studies involving the use of augmentative and alternative communication (AAC) strategies. The participants were a 59 year old female, MC, and a 60 year old male, CE, both of whom had been suffering from progressing language decline for 5+ years prior to the initiation of the study, and were otherwise physically

healthy individuals. A communication needs assessment was undertaken for both MC and CE. Both patients had generated their own strategies for communication, and these were incorporated into the aids. MC successfully used augmented input (i.e. use of writing out words, visual cuing). No follow up information on her continued success was available, due to her distance from the assessment site. CE participated in a month of intensive AAC development, training and application. These focused on cued comprehension and augmented expression in routine and unfamiliar contexts. He received three to four hours of direct therapist training sessions on these techniques per week as well as communication at home and in the community facilitated by the therapist. His family was trained on both receptive and expressive communication strategies that would be helpful when communicating with CE. Data was recorded from the family via tape recordings of conversations and informal notes and impressions, though these did not give significant information on non-verbal communication. The therapist and communication partners also kept a tally of when symbols had been introduced and were perceived to be understood. At baseline, CE did not appear to understand any of the symbols, but by the third session, CE was able to spontaneously use symbols appropriately, and demonstrated comprehension of 70% of symbols, half with gestural or contextual cues. His family reported observing increased comprehension, and more complex expression, along with increased turns when CE used his communication boards. In a follow-up session with CE a year later, CE's language difficulties had progressed substantially, and he now had little to no comprehension of verbal communication and relied heavily on gesture and sound effects to convey his message. Family reported that he was still able to use his self generated AAC book and communication boards to increase his communicative effectiveness.

Both of these patients achieved success with the use of AAC devices, particularly those which capitalized on use of the preserved skills as well as those that matched self generated strategies. This study has relied heavily on qualitative data, and while it is very suggestive of the effectiveness of using AAC strategies with people with PPA, further studies need to be conducted to make a conclusive statement. A further concern is the lack of blinding in this study, as experimenter bias could be very prominent, as much of the data was collected by family members and the therapist working with CE knew him socially prior to commencement of the study.

Pattee et al(2006) report a case study in which they compared two AAC strategies on communicative effectiveness in an alternating treatment design. The two strategies are training the participant on a text-to-speech device (ACD), the LightWriter (Toby Churchill, Cambridge, UK) and American Sign Language (ASL). The participant, a 57 year old female, presented with severe Apraxia of Speech (AOS) and impaired expressive communication as the disease progressed. Her receptive communication was within functional limits on formal testing. The study was conducted over a period of 9 weeks with two baseline data collection sessions, 8 treatment sessions and three post-treatment data collection sessions. Output was transcribed and analyzed using an adapted form of the Nicholas and Brookshire (1993) correct information unit (CIU) protocol. In both modes, treatment sessions consisted of the subject being asked to describe a photograph card of a daily activity, with a focus on eliciting phrases with agent, action and object, and 'Wh' concepts (who, what, etc.). Directly after each treatment session, a novel photograph was presented. Once a week, repeat probes were administered at the beginning of the session. Post-intervention analysis of the CIUs in the transcripts revealed an increase of 2% in correct CIUs for the ACD and an increase of 24% for ASL. For the ACD, maintenance of 93% correct CIUs was maintained for the repeat probes, and an increase from 20-85% was noted for the ASL condition. Novel probes revealed a slight increase in correct CIUs for both conditions.

While these results may seem striking, the authors acknowledge that the patient's familiarity with the English writing system, as it is her first language, versus her relative unfamiliarity to ASL, may have confounded the results. Of note, the subject preferred the use of ASL to the ACD, even though it would not allow her to communicate in as wide a context. Both strategies were helpful with increasing the subject's communicative effectiveness, and patient preference must be taken into consideration when choosing an AAC strategy. A treatment design that compared CIU at baseline without the strategy and then after training on the strategy may have been more elucidating, as it would have allowed us to see which facilitated more effective communication in comparison to the same baseline (no aid). Further studies may want to consider this approach. Overall, both treatments improved her communication and may be considered successful, though more research must be conducted to make a more conclusive statement.

#### Other Treatment Protocols

Finocchiaro et al (2006) present a case study in which they trialed the use of high frequency repetitive Transcranial Magnetic Stimulation (hf-rTMS) in improving the patient's linguistic skills. Transcranial Magnetic Stimulation (TMS) is a non-invasive procedure which is hypothesized to normalize the neural activity in cortical areas of metabolic dysfunction. hf-rTMS has been shown to increase the cortical excitability of the area (Pascual-Leone et al, 1998). The participant is a 60 year old man, whose MRI scans showed atrophy predominantly in the left fronto-temporal region. His linguistic skills were below normal limits on both formal and informal tests. Further analysis showed he had an increased impairment of verb naming over noun naming. The hf-rTMS was applied to the left prefrontal cortex, in half hour treatment blocks, and the patient's performance on language production (sentence completion) and memory span tasks was evaluated both before and after two 5 day hf-rTMS treatments and one 5 day SHAM treatment, in an alternating design, with a return to baseline between each condition. The tester was blinded to the hypothesis at the time of testing. The experimental task was administered two times to obtain baseline data, in order to control for learning effect, as the entirety of the test was administered at each test point (total of 12 times). Testing was conducted one to two days post hf-rTMS treatment/SHAM, and no stimulation occurred during this time. Repeated measures ANOVA were performed on the scores for each task. The two main variables of Session and Task were present in the sentence completion task, while the only variable in the memory task was Session. The verb sentence completion task was significant for the effect of Session, ( $F(12, 408) = 3.3, p = .0002$ ), due to the increased performance after both hf-rTMS treatments. Duncan post hoc comparisons also revealed significant differences between scores after the treatment versus baseline data and versus post SHAM scores. The only significant Task effect was for the noun exercise, which revealed that the participant was much better at the determiner task regardless of treatment. No variables were significant for the memory tasks.

This is a well designed case study that shows hf-rTMS may have an effect on verb finding in a sentence completion task. It has been well controlled with an alternating design, including a control for the return to baseline between conditions tested, and blinding. While the authors have controlled for learning via the repeated administration of tests to gather baseline data, the repeated use of the test multiple times in a relatively short time span is

concerning. Because the entire test was administered numerous times, and part of the profile of PPA is lack of cognitive decline, learning/memorizing of the tasks may have occurred to some degree. While this study has reported some interesting and suggestive findings, additional rTMS studies are required to strengthen this efficacy evidence.

McNeil et al (1995) present a hybrid single subject experimental design, with components of multiple baseline, multiple probe, and withdrawal designs. A dual treatment protocol, consisting of behavioural and pharmacological treatment, was used. The subject was a 61 year old male, GP, who presented with a mild linguistic processing deficit, with no deficits in other areas of processing and a rapidly progressing spastic dysarthria. Within the design of the study, multiple baseline sessions were conducted to assess GP's performance on a list of several potential treatment targets as well as standardized speech and language tests and connected speech samples. Subjects were asked to produce either synonyms or antonyms for predicative adjectives, with the application of a cuing hierarchy when necessary to facilitate word retrieval. The pharmacological treatment, dextroamphetamine, was administered daily. Data was collected from both behavioral only and behavioural plus pharmacological phases of the study. Results showed an increase in ability to name the treated items as well as generalization to other words and word classes (verbs and prepositions), with no difference in effect between the behavioural only and behavioural plus pharmacological treatment conditions. Thus, there is poor evidence for the use of dextromamphetamine.

While this study provides persuasive evidence that this treatment paradigm is effective for people with PPA, there are some methodological concerns. The outcomes were judged by two experts not affiliated with the study, who visually examined graphs presented of the data collected during the sessions. While the data in the graphs is well set up and shows gains in the GPs performance, the lack of any analysis other than visual reduces the validity of the design. Also, due to GP's performance above criterion at baseline, nouns were taken out of the design and this early termination of data collection weakens the experimental design. Finally, it is a single subject design and thus has limited generalizability to the population. However, the fact that these gains were seen against a backdrop of linguistic decline in other areas does make a strong argument for treatment. Overall, this is a relatively well designed study, which, while it should be

interpreted cautiously, does make a good argument for treatment.

Rogalski and Edmonds (2008) present a case study in which treatment for PPA is aimed at the discourse level. The treatment protocol used was Attentive Reading and Constrained Summarization (ARCS), a novel discourse level treatment derived from cognitive principles, designed to operate on both macro- and micro- structure linguistic skills. It is based on the assumptions that attention is important for complex language and that intentional language use promotes language generalization by encouraging verbal language to the exclusion of other modalities. The subject is a 76 year old male with diagnosed PPA and concomitant attention deficits. His discourse level communication and naming abilities were impaired, while his auditory comprehension and repetition were relatively preserved. Treatment consisted of having the subject read through news articles and summarize content as he read. He was asked to summarize, with the constraints of (a) no opinion on the passage, (b) no use of pronouns, (c) no use of non-specific words. Coherence, cohesion and informativeness analyses were performed on narrative pre- and post- treatment and two month post language samples. Results showed a substantial increase in all areas post-treatment, with the exception of words per minute (used as a measure of informativeness). These gains were seen in maintenance measures also, with measures increasing or remaining above baseline. This may in part be due to the subject continuing to practice on his own.

This is a well designed study, which was based on clear theoretical concepts. The results related to the effectiveness of ARCS as a treatment for PPA are suggestive, though results must be interpreted with caution due to its nature as a single case study. The measurements used have high construct validity, as they are strongly related to discourse. The positive maintenance measures also increase the strength of evidence for this treatment, though they could be confounded by the participant's continued practice post-treatment. Further research in this area is warranted.

Murray (1998) presented a longitudinal case study of a patient, DD, a 64 year old woman with PPA. She describes the different therapies and their outcomes across the progression of the disorder. Longitudinal speech and language testing showed a decline across modalities, with her verbal expression being most severely affected and auditory comprehension declines appearing in later stages. Reading and

writing were relative strengths and she was able to maintain fairly effective functional communication.

The first treatment approach was a traditional stimulus response therapy, focusing on auditory and reading comprehension and spoken and written expression. Positive outcomes were seen, however, DD exhibited very little carryover and was perceived as having much deteriorated verbal expression, preferring to use writing. Thus, this treatment protocol has low ecological and construct validity and the results must be interpreted very cautiously.

The second treatment protocol, capitalized on DD's relatively maintained written communication. 'Back to the drawing board' (BDB) (Morgan and Helm-Estabrooks, 1987), a drawing program which seeks to capitalize on upon intact right hemisphere function and possibly deblock verbal and written expression was used. Naive judges rated pre- and post- treatment drawings. Pairwise t-tests were undertaken and showed that the pre- and post-treatment results were only significant ( $p < 0.05$ ) for drawings of two part scenes. DD also showed an increase from 67% to 83% on the WAB Drawing subtest. Non-treated domains remained stable, and thus the improvement in drawing may be a sign of treatment effect. Little carryover effect was seen, though this may be confounded by the fact that the writing was an easier means of cuing in the generalization tasks. This therapy was terminated in favour of functional communication training with both DD and her spouse, as at this point, frequent breakdowns of communication were occurring between DD and her spouse and negative interactions were observed. Qualitative analysis of pre- and post- treatment showed that the couple used more effective communication and repair strategies after treatment. Furthermore, both individuals were observed to use a greater number of communication modalities, including DD spontaneously using drawing, 4 months after the BDB therapy had been terminated. Functional communication measures remained relatively stable across time, while linguistic skill continued to deteriorate, suggesting positive treatment outcomes.

This longitudinal study makes a strong argument for speech-language pathology intervention across the progression of a client's disorder. It lays a foundation for future longitudinal research to follow.

### ***Recommendations***

The above studies all show some evidence that speech language therapy is warranted for individuals

with PPA. Because they mainly include only single subjects, they have inherent external validity concerns for generalizability, especially as this population is very heterogeneous in their linguistic profiles. From the above studies, recommendations for future studies, as well as for clinical practice, can be made.

### ***Future Research Implications***

- Single subject multiple baseline experimental designs provide a higher level of evidence than case studies, and should be used in situations where only one subject is available
- Where possible, multiple subject designs should be conducted to increase external validity
- Post-treatment studies should include testing of linguistic processes not likely to be affected by treatment, to allow comparison of general linguistic decline versus decline in the treated areas. This would allow more valid measures of effect in the context of a progressive disorder.
- Sufficient data regarding the procedure used must be included to allow the study to be replicated, thus improving the evidence base for a particular treatment.

### ***Clinical Implications***

- Functional communication strategies and training should be considered for patients with PPA and their communication partners, with recognition of the fact that it is a progressive disorder, and that there may be very little cognitive impact outside of language.
- The evidence for use of AAC strategies is compelling for this population. Use of ACC devices should be initiated early in the progression of the disorder to capitalize on their present skills to learn and personalize the device.
- rTMS as a treatment for PPA has a moderate level of evidence, but requires further testing to improve validity.
- Treatments designed to target a specific linguistic structure/process have a suggestive evidence base, but show little generalizability to naturalistic contexts.

- Increasing the number of modalities in which the patient can communicate appears to have a positive influence on their communication, though more research is necessary.
- Treatment should be made as naturalistic as possible, to facilitate carryover, as this was a common issue seen in the studies

### **Conclusions**

There is persuasive evidence that individuals with PPA can benefit from speech and language treatment. However, further efficacy research is necessary.

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