

Primary Progressive Aphasia: The Journey to Diagnosis and What We Know about Treatment for Communication Difficulties

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Journey to Diagnosis

Receiving a diagnosis of primary progressive aphasia is hard. When communication starts to change it often starts off subtly – forgetting a word every so often, speech rate and voice may sound a little different, processing of information might seem a little slower. Sometimes it may only be in hindsight that all of the early signs of difficulty even become apparent.

These early stages, even before diagnosis, might be some of the most challenging. A lot of people report confusion – they don't know why they or their loved one are doing what they're doing. There can be impatience and tension because loved ones don't know how to handle the situation, or how to best help. Within medical settings, there might even be some symptoms or behaviors that get written off as insignificant or mislabeled as some other problem (e.g., depression, stress, etc). Further, because people with primary progressive aphasia start to have communication difficulties in late middle-life, it can mean that a progressive condition isn't even considered. Isn't that something that typically happens to older people? On top of that, it may often take a few years and multiple referrals to get the right diagnosis, eventual loss of work may place financial burdens on families, and changing roles can make the person with primary progressive aphasia feel dependent on others.

All of this is to say, that as a clinician, I recognize that this is complicated. That each person and each journey to diagnosis is different. As someone who has not had a loved one with primary progressive aphasia, I do not know all of the ins and outs of what it took for a person and their family to end up seeking help from me. But what I do know, is that the best first step, with any person, is to listen. To find out what it is that makes them them, to find out what is challenging, and to find out how I can best help.

With most speech and language therapies, it is important to know that there is not a cure. I wish more than anything that we could make everything go away and for the person to be “back to who they were before”. But saying that there isn't a cure, and that there isn't hope are two different things. There are activities, skills, and behaviors that we do know can help people with primary progressive aphasia and their loved ones.



What We Know about Treatment for Communication Disorders in Primary Progressive Aphasia

The primary goal of speech-language treatment for people with communication disorders due to primary progressive aphasia is to maximize communication. Maximizing communication, though, doesn't mean correcting or achieving perfection within talking, understanding, reading, and writing. It means finding what personalized approaches can help to support communication and improve engagement and participation in desired life activities. Most often, this is best achieved by a multi-component approach.

One of the first arms of treatment is to target specific speech or language skills that are challenging for the person with primary progressive aphasia. The specific targeted skill depends on the person and how far along the disease is, but a common difficulty is coming up with the names of things in conversation. Research has looked at a person with primary progressive aphasia's ability to name a set of words after treatment. Sure enough, the person is able to more accurately and effectively come up with the trained names immediately after treatment and, for some studies that gather long-term data, the effect lasts for up to six months and one year later. Other studies even show that the effects of treatment generalize – the naming improvement can extend to words that weren't directly trained. However, this usually depends upon how similar the words are (either based upon their meanings and/or their sounds) to the trained words and isn't observed in every study. Other tasks besides naming can be trained, too. For example, learning to more fluidly and efficiently produce speech in trained "packets" of content (e.g., talking about what you used to do for work, describing a favorite vacation, ordering from a restaurant). Overall, the main theme for this first arm of treatment is that training can help improve performance on specific tasks. Like I said, it isn't a cure, but it can certainly help.

Another important arm of treatment is training strategies and supports. This might include identifying strategies that you and your family have already found to be beneficial and continuing to encourage their use. Or it may mean finding behaviors that aren't helpful and replacing them with more facilitative ones. The speech-language pathologist that you are working with should also explore supports for communication as part of this arm of treatment. That could include picture supports, read aloud features, adjusting font size and spacing on documents, eliminating background noise, or using reminders. This list is not exhaustive, though. Often this arm of treatment requires creativity and accurate identification of the barriers within everyday tasks. But when the appropriate strategies and supports are identified and trained they can help a person with primary progressive aphasia regain modified independence with tasks and feel more confident and successful. Ideally, these strategies and supports are also implemented preemptively – so that training can be taught and engrained early before the task becomes too difficult or before the cognitive effort of learning the strategies and supports becomes too challenging.



A third important arm is communication partner training. Communication is the exchange of ideas and information between two people. So, ultimately, in order to improve communication, it often means adaptation and flexibility from the communication partner (e.g., spouse, friend, or child of the person with primary progressive aphasia). Changing a behavior or a response to a behavior takes time, though. It is like forming a new habit. First you have to identify what you want to do or change, consciously target that behavior, practice it over and over, and then over time, it becomes more automatic. It's natural for this to feel unnatural at first – and that's why it can be beneficial to tackle in treatment so that the speech-language pathologist can provide guidance and feedback.

A final arm of treatment is education. Primary progressive aphasia is a rare and complex disorder. There are different diseases that cause the different subtypes of primary progressive aphasia, and the terminology can be confusing. It helps to have someone explain this information, answer questions, make referrals to other providers, and ultimately help plan for the future as able. However, it is also important to recognize that this can and should be done in a personalized way as well. Depending on the person and their family, their journey with coping, other life events at the time, and a myriad of other variables, information at point A may not be as helpful as information provided at point B. So, it is part of the therapeutic alliance between the clinician, the client, and their family to best navigate the timing and amount of education that is provided in treatment.

Primary progressive aphasia is a complex and challenging disorder. It is not something that many people are aware of, and certainly not something that families plan to face before it happens. But, my hope is that through an empathetic and well-rounded treatment plan, that speech-language pathologists can empower the person with primary progressive aphasia and their family to maximize communication and to get the most out of every interaction.

About the Author



Miranda Babiak is a speech-language pathologist at the Pittsburgh VA working in their intensive aphasia treatment program (PIRATE). Previously, she taught and supervised graduate students in speech-language pathology at the University of Northern Colorado. She received her Clinical Doctorate in Medical Speech-Language Pathology from the University of Pittsburgh in 2016. Prior to that she worked in research at the University of California San Francisco. There, she worked extensively with leading researchers who are trying to better understand primary progressive aphasia (PPA), treatments for PPA, and the impact of PPA on the person and their family. She also worked with people with brain tumors and intractable epilepsy. Her primary goal in her career has been to better understand what happens when communication breaks down and to find the best ways to help and support the person and their family who experience these breakdowns in their everyday life.

