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The Other PPA

Primary Progressive Aphasia

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For those involved with Positive Approach® to Care (PAC) the acronym for Positive Physical Approach™ is PPA™. However, to many neurologists and speech-language pathologists PPA means something entirely different, namely: Primary Progressive Aphasia.

Primary Progressive Aphasia (PPA) in Brief:

- First described by Dr. Arnold Pick and Dr. Paul Sérier in 1892-93
- Framed as primary progressive aphasia in the 1980s by Dr. Marsel Mesulam
- Uncommon syndrome primarily affecting language
- Insidious progressive language impairment is primary deficit for two years after onset
- Minimal deficits in other cognitive domains (memory, executive functioning, etc.)
- PPA has three major variants:
 - (a) logopenic – missing words
 - (b) semantic – missing meaning
 - (c) nonfluent – missing structure

PPA – A Look At Language

Mary calls out: *“John! How are you?”*

“Good,” he replies. “How are you?”

John knows what he wants to say. And he is physically able to produce the words. He is able to speak clearly, and the words come out in the correct order. John does well with simple chit-chat.

Mary then asks, *“John, you were a mechanic for 30 years? My car is leaking oil. What do you think is going on?”*

John is happy to be asked. He loved being a mechanic and never really wanted to retire. *“Well, it could be the oil panel or the...”* John pauses, looking for the word he wants. *“the oil, or the transmiss...”* he hesitates. *“Or it could be the...you know, the connector...the gadget.”*

“Oh, I’m not good with gadgets.” Mary is starting to get discouraged.

“No, not gadgets, the... you know, the seating, the black rubber round...you know, the daskets.”

“Daskets!?” Mary sighs. *“Can you take a look at it?”*

“Sure, let me get my tools.” And off they go.

John has a form of primary progressive aphasia known as logopenic PPA. Logo comes from Greek (*lógos*), meaning “word,” and “penia” means deficiency. Logopenic then indicates a deficiency of words.

Let’s look at where John ran into trouble with his words in the example above. John was trying to say that Mary’s oil leak could be the result of a bad oil pan or a faulty gasket.

When he tried to say oil pan, it came out “oil panel.” The phonological speech errors involved include substituting alternate words for the desired words and trailing off and not pronouncing the final consonants. John did this when he said “transmiss..” rather than transmission.ⁱ

People with IvPPA or PPA-L (logopenic primary progressive aphasia) will often experience phonological paraphasia -- substituting a sound-alike word-form for the word they wish to use. John was trying to say “gasket” but it came out “dasket.”

At one point, when John couldn’t come up with the word “gasket” he talked around it, trying to describe it: “you know, the seating, the black rubber round.” This is called circumlocution and is common with people living with logopenic PPA.

Finally, notice that when the conversation was just chit-chat, John’s speech was fluent and showed no signs of impairment. It was only when he was trying to express complex concepts that he had trouble with word-finding and properly expressing the words he wanted to use.

People with logopenic primary progressive aphasia usually know what they want to say but have trouble expressing it. Speech can be fluid and grammatically correct but marked by difficulty in finding words (which can cause hesitation). Other common features include using the wrong words, not fully pronouncing words, and talking around or describing what is wished to be named.

This is a progressive condition. As it gets worse, the person will have more difficulty producing and comprehending complex sentences.

When John goes to his neurologist, Dr. Smith gives him a complex sentence to repeat back to him. Each visit, this becomes harder for John to achieve and he makes more and more errors.

Dr. Smith explains to John’s family that his cognitive window is becoming narrower and giving John complex instructions will become harder for him to follow. John’s ability to understand words and what they mean is largely retained but stringing together complex thoughts and compound language will be more difficult for him in the future.

John knew what he wanted to say but had trouble producing the correct words. His errors occurred with the formation of the words and choosing the right words rather than the order of the words or knowing what the words mean.

Ted also goes to Dr. Smith, a board-certified neurologist specializing in neurodegenerative diseases. At the last visit, Ted could speak fluently and repeated back the complex sentence that John had trouble completing. Nonetheless, Ted is having trouble following instructions. Last night when helping his wife bake biscuits for their twin grandkids, Carolyn got angry when he brought her a frying pan. She yelled at him: "Does that look like a Rolling Pin?"

When talking privately to Dr. Smith, Carolyn confessed: "Doc, I feel so bad. I just lost it. I don't know why he's doing this. He knows better."

"Maybe not," Dr. Smith responded. "Ted has semantic primary progressive aphasia, or semantic dementia. It falls under the class of Frontotemporal dementia. This means the front of his brain is affected. This is where we make decisions. And the temporal lobe is also affected. This is involved in hearing, processing of language, and ability to speak." Dr. Smith motions to his temples to point out where the right and left portions of the temporal lobe are located.

"When you told him to bring you a rolling-pin, his brain had to hear what you said -- not just his ears -- but his brain. The temporal lobe is what allows us to process all the sounds our ears pick up and make sense of it all. He has damage to this part of his brain."

"So, he didn't even hear what I said?" Carolyn really wanted to understand.

"Yes, but not because of his ears. Because of his brain. This is not a hearing aid issue. Speaking louder will not correct this. Think about what needs to happen for him to bring you a rolling pin."

"First, he needs to hear the words. Next, his brain (in the temporal lobe) needs to process what was said and attach meaning to it. The words are tied to ideas and concepts and oftentimes to images. After that is done, Ted needs to go find the rolling pin, pick it up, and bring it back to you."

"That simple little instruction is quite complicated. It demands that his brain is working well in the front where decisions are being made (where to look), in the temporal lobe where auditory and other sensory information is being processed (to hear and understand the words spoken), in the cerebellum to coordinate movement and balance (to walk, bend down, and move around), and in the occipital lobe that processes visual information (to be able to see what to grab and bring it to you)."

"I can't tell you what went wrong or where a breakdown occurred in his cognitive processing that caused Ted to come back with a frying pan rather than the rolling pin, but this is to be expected with semantic primary progressive aphasia. It's called semantic primary progressive aphasia because 'semantic' means relating to the meaning of

language or words. That's what he's losing, the ability to name things and know what they are."

"It sure can be frustrating. Maybe you can tell yourself that he's not doing any of this on purpose. He was trying to help. Somewhere along the way, his brain told him that you wanted a frying pan and that's what he got you. It could be because his semantic PPA caused him to attach a different meaning to the word rolling pin or could be because when he saw the frying pan he just thought that's what you wanted. We'll probably never know. But it seems clear that he was trying to help, and it seems clear you love him a lot."

"Oh, I do Doc." She shuffled through her purse to find a tissue. "What can I do? Should I just stop asking him to help me?"

"What you can do is forgive yourself. You have a long way to go. You need to be kind to yourself. Beating yourself up does you no good. And does him no good."

"I know." Carolyn wiped the tears from her eyes. "I know."

"Good. And you want him helping. I bet he's a good helper."

"He is." Carolyn smiled. "He's a good man."

"How about showing him a picture of a rolling pin? Or pointing out where you keep it and describing it rather than just labeling it?"

"I could do that."

"Great. How about thanking him for bringing you the frying pan when you asked for the rolling pin? Can you do that?"

"I'll work on it."

People with semantic primary progressive aphasia often have trouble naming and recognizing everyday objects (anomia). They will have difficulty reading and writing words that are not spelled phonically (dysgraphia). As the disease progresses, it can be difficult to understand what someone living with semantic primary progressive aphasia is saying because words are omitted, or the wrong words are substituted.ⁱⁱ

A major difference between logopenic PPA and semantic PPA is the ability to understand what words mean. In general terms, people with logopenic PPA (lvPPA or PPA-L) will know the words they want to use but often have trouble producing them. People with semantic PPA (svPPA or PPA-S) will often not know what words and objects mean, even if they can pronounce them with fluent speech.

Besides these two variants of PPA, there is a third: agrammatic or nonfluent PPA. Two key elements distinguish this form of PPA from the other two types discussed above: apraxia and nonfluency.

As the name implies, non-fluent or agrammatic PPA is characterized by speech that is halting, broken up, or otherwise mixed up. Words are used in the wrong order. The wrong verb tenses are used and many words (such as pronouns, conjunctions, and articles) are omitted. This creates speech patterns that appear disorganized and difficult to follow.

In addition, the ability to form sounds is compromised. This is called apraxia of speech. As related to the non-fluent variant of primary progressive aphasia (nfvPPA or PPA-G), the muscles needed to produce speech are themselves unaffected. However, the functions of the brain and the network of neurons required to produce speech are damaged by nfvPPA. In essence, the brain is not able to successfully tell the lips and tongue how to move to produce the desired words.

People living with progressive non-fluent aphasia will often have labored speech. Speech is usually slow, distorted, and there are sometimes exaggerated facial movements that accompany the effort to speak.

Review

Just as dementia is a term given to a category of different but related diseases, likewise PPA is not a single disease or condition. There are commonalities that join the three major forms together and there are distinctions between them. Historically, the different forms of PPA have been described as subsets or variants of frontotemporal lobar degeneration (also known as frontotemporal dementia and formerly called Pick's disease). But not all cases of PPA are related to frontotemporal lobar degeneration. Recent research has shown a stronger connection between logopenic PPA and Alzheimers disease.

Semantic PPA is almost always related to frontotemporal lobar degeneration (FTLD). About three-quarters of cases of non-fluent PPA are related to frontotemporal lobar degeneration.

Logopenic PPA is related to Alzheimers more than FTLD or other disease processes.

Recap Reminders

Logopenic: (word deficiency) the word is missing, but the meaning is there.

Semantic: (meaning of words) the meaning is missing, but often words can be read, said, and repeated.

Nonfluent: PPA results in speech that is broken up, not in the correct order. With nonfluent PPA the structure and pattern are missing, but the meanings of words are retained.

Clarke Pollard currently lives in Naples, Florida. He joined PAC in 2017 and currently serves as the Executive Director of the [Alzheimer's Support Network](#), a 501(c)3 Nonprofit that offers private consultations, support groups, respite programs, [Wanderer's Identification program](#) (Collier County Florida), a 24-hour helpline, and more.

Pollard states, “the names of the persons presented in the article have been changed. Their stories are used with permission. Dr. Smith is an amalgamation. Quotations in this article are not attributable to a single person. Quotes are presented for presentation purposes. Thank you to all who assisted. We are honored that Teepa serves as a Member of the Board of Directors for the Alzheimer’s Support Network.”

*Feel free to contact [**Clarke Pollard**](#) with comments, suggestions, or corrections.*

ⁱ [Logopenic Variant PPA](#). (n.d.). Retrieved May 1, 2018.

ⁱⁱ [Semantic Variant PPA](#). (n.d.). Retrieved May 1, 2018.

ⁱⁱⁱ Harris, J. M., Gall, C., Thompson, J. C., Richardson, A. M., Neary, D., Plessis, D. D., Jones, M. (2013, November 19). [Classification and pathology of primary progressive aphasia](#)

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