

When the Meaning is Lost - Semantic Variant PPA

There are several forms of primary progressive aphasia (PPA). Recognition of differences and careful evaluation are important for care planning. Individuals with semantic variant PPA lose the meaning (or semantics) of words, have trouble recognizing faces of familiar people and have difficulty understanding the emotions of others. In addition disinhibited, rigid and compulsive behaviors eventually develop. While most remain relatively unconcerned about their condition, some individuals retain insight and focus on their deficits and may be at risk of suicide.

Meet Betty James

Mrs. James was a 68-year-old divorced, college graduate who retired from her job as an executive secretary at age 65. She lived alone in a one-bedroom condominium. Her one adult daughter resided six blocks away and was concerned about her mother's slowly worsening "memory" problem, which she described as forgetting people's names, trouble remembering words, and difficulty recognizing friends and her twin brother whom she saw several times a year. Mrs. James didn't seem to "care" about her two grandchildren anymore. Instead of visiting when they returned from school, she now chose to nap from 1-2 p.m. and then played computer solitaire all afternoon.

Mrs. James agreed she was having some trouble with words but did not appear concerned. When asked why she no longer spent time with her grandchildren, she stated naps were important, as one got older. She was very careful about her diet and stopped eating at restaurants because she believed she had multiple food allergies. Mrs. James shopped for groceries, prepared simple meals, paid her bills on time, volunteered weekly at a community hospital and sang in her church choir. She took the local bus to these activities. During the initial evaluation, Mrs. James was shown pictures of 15 common animals and objects and could only name five of them correctly, often responding, "What is it?" She correctly copied three-dimensional objects indicating intact visuospatial domains. She adequately performed simple arithmetic calculations.

Diagnostic brain scans including both a magnetic resonance image (MRI) and positron emission tomography (PET) revealed marked anterior temporal lobe atrophy, with the left lobe more affected than the right. The clinical history, behavioral issues, cognitive evaluation and imaging results were consistent with a diagnosis of semantic variant PPA.

Worsening Symptoms

Over the next 18 months, Mrs. James' ability to name and use objects continued to worsen. She no longer rode the bus after becoming confused about showing her bus pass to the driver. She continued to attend weekly choir practices and church services. She became more rigid in maintaining set routines, particularly in her diet. She carried a list of food she could eat and insisted upon checking the list before putting anything in her mouth, although she was unable to describe what the words meant. Her daily diet included: Cheerios cereal, banana and milk for breakfast; peanut butter and jelly sandwich for lunch; and Jenny Craig spaghetti microwave dinner. She became increasingly conscious of time and tapped her

New Email Group

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Did You Know?

Primary progressive aphasia is a clinical dementia syndrome. Degenerative disease causes language abilities to slowly decline and over time affects other cognitive and behavioral skills. Symptoms include: word finding, anomia, speech errors and poor auditory comprehension. There are three PPA subtypes: nonfluent/agrammatic, semantic and logopenic.

PPA is diagnosed through a series of exams: neurological to determine the rate of onset, and other possible illness; neuroimaging to rule out other diseases; neuropsychological to determine the nature of language impairment; and speech-language pathology to assess language modalities (speech, reading, writing), components (grammar, semantics, phonology) and functional communication skills for treatment planning.

FTD and PPA Conference

Northwestern University FTD and PPA Conference – Monday, November 5th in Chicago. CEUs are available. Visit the conference site for more information: <http://www.brain.northwestern.edu/about/events/ftdppa.html>



watch constantly on the way to church stating “I need to go in 10 minutes, now nine minutes,” etc. Her daughter moved her to a small memory care facility after discovering Mrs. James bought cat food (instead of cereal) and ate coffee grounds directly from the tin.

Creative Interventions Ease Transition

During the first few weeks in the facility, Mrs. James initially refused to take a shower and change her clothes, asking, “What’s a shower?” She would not eat, as the foods were “not on the list.” She preferred to sit alone in her room. Her daughter completed AFTD’s [Daily Care Snapshot Tool](#). She met with the facility interdisciplinary team to discuss Mrs. James’ preferred times for activities and ways to incorporate her need for routine in planning care. A whiteboard was placed on Mrs. James’ bathroom door with a picture of a shower and clock set at 7 a.m., her preferred showering time. Her daily schedule was also posted on the whiteboard. Staff greeted her in the morning, pointed to her watch, then said, “It’s 7 o’clock,” pointed to the picture on the bathroom door and guided her to the shower. While she was showering, staff replaced her dirty clothes with clean ones.

A separate buffet table with her name on a placard was set up in the dining room with her usual brands of food for each meal. After several weeks, other foods were added, which

she occasionally ate. She went on morning walks with other residents. She ate lunch at noon, took a nap, and then played computer solitaire in the game room. After dinner, she listened to music with other residents and enjoyed singing along to favorite tunes. After an adjustment period, she settled into this routine and remained in good spirits. Mrs. James smiled and said “hello” to other residents and staff but otherwise did not converse with them. She continued to go to church on Sundays with her daughter’s family but insisted on returning to the facility to eat at her own table.

Mrs. James began spitting instead of swallowing what was in her mouth. She did not seem to recognize the meaning of saliva, and she thought it would make her sick. Her family no longer took her to church with them because of the spitting. Other residents in the facility were understandably upset with this compulsive behavior. When she began spitting in the dining room and other common areas, she was asked to leave the program. After physical and dental examinations did not reveal a medical cause for the spitting behavior, Mrs. James was prescribed an antidepressant that decreased the frequency of the spitting, but did not resolve it completely. In the most recent meeting with her daughter and staff, Mrs. James’ care needs were discussed. In deference to other residents, Mrs. James continued to choose her food from “her” table, but ate in her room in the

Troubles & Tips

Q: *We work with a man who has semantic PPA and has become depressed and hopeless. Is there a risk of suicide among people with this diagnosis?*

A: Yes. Many people with semantic PPA are not bothered by their losses. However, a percentage of people (possibly 20%) are aware and troubled by the losses.

Semantic PPA typically affects people at the prime of their career and family life. The symptoms begin gradually with difficulty finding the right word or the names of friends. People are able to speak fluently, but lose the substance of conversation as the meaning of more and more words is lost. Many people have trouble recognizing faces and reading emotion or non-verbal communication while visuospatial skills and the ability to do calculations remain preserved.

The loss of understanding words and concepts makes it difficult to participate in activities and relationships that have been important to the person. People become unable to work and are often granted early social security disability. Roles at home change as a spouse continues or returns to work to compensate for lost income. The “invisible” nature of the cognitive deficits means family and friends may not recognize the changes as symptoms

of disease and become embarrassed or impatient when with the person.

As the semantics of language are lost, hobbies become impossible. For example, someone who did woodworking projects or enjoyed cooking and entertaining can no longer recognize the tools or utensils needed, or a sports fan no longer understands what players on the court are doing. Over time, the details of occupations, behaviors, and feelings a person experienced in the past become more vague and devoid of meaning. For example, one person could state he was a “father,” but was unable to describe his son as a baby or how it felt when he was born, and could not imagine his son with a family of his own.

People with semantic PPA can become despondent about how “stupid” they feel and may recognize an increasing burden on the family. Emerging research indicates that sv-PPA patients who had difficulty projecting themselves into the future and had disinhibited behavior were at greater risk. In light of the lack of pharmaceutical treatment for PPA, particular attention must be paid to psychosocial support and intervention. Should a person begin to express feelings of hopelessness or that their family would be better off without them, the risk of suicide should be evaluated. Antidepressant medication may be considered, and safety precautions, such as one-on-one coverage, could be implemented round the clock until staff assess that the risk is gone.



presence of a staff member. She also played computer solitaire in her room rather than the game room with others. She was able to participate in singing with other residents and did not spit during this time. Her spontaneous verbal output continued to decrease and she was able to shower and eat with assistance. She became less active, was at risk for constipation and developed urinary incontinence.

Questions about the case:

1. What signs and symptoms indicated that Mrs. James was losing the semantics or the meaning of words and concepts?

Mrs. James began “forgetting” people’s names, had trouble remembering words, and experienced difficulty recognizing friends and her twin brother. During her initial evaluation, she was shown pictures of 15 common animals and objects and only named five of them correctly. Her response to several, “What is it?” was distinct from a response such as, “I can’t remember.” Her visuospatial domains remained intact, so the problem was not in processing the image visually, and she could perform simple arithmetic calculations. Mrs. James carried a list of foods she could eat, but was unable to describe what the words meant. Later she bought cat food (instead of cereal) and ate coffee grounds directly from the tin.

2. What additional behaviors developed that presented challenges for the facility staff and how did they respond?

Mrs. James refused to take a shower and change her clothes, asking, “What’s a shower?” Staff read her Daily Care Snapshot and met with her daughter to incorporate her preferences, needs and routine into her care. Shower time was scheduled at her preferred time, 7 a.m. A whiteboard with picture of a shower, clock set at 7 a.m., and her daily schedule was placed on her bathroom door. Staff pointed to her watch and the picture of the shower, provided a verbal cue that it was 7 a.m. and guided her to the shower. Dirty clothes were replaced with clean ones while she showered.

Mrs. James refused to eat, as the foods were “not on the list.” Rather than try to change her rigid, compulsive behavior, a separate buffet table with her name on a placard was set up in the dining room. She was served her usual brands of food. Seeing her name and familiar foods eased the transition and slowly, other foods were added.

Mrs. James preferred to sit alone in her room. As the disease progressed, she began to engage with others less, likely because it became harder to understand conversation. Staff invited and escorted her to activities that the daughter identified as interests. These included walks, playing computer solitaire, and listening and singing familiar tunes. An afternoon nap was scheduled daily and a new routine was established.

Mrs. James began spitting instead of swallowing what was in her mouth. She did not seem to recognize the meaning of saliva. Physical and dental exams were completed. An antidepressant was prescribed to decrease the frequency of spitting. When she began spitting in common areas, she was escorted from those areas. She ate (with staff supervision) and played computer solitaire in her room. She participated in group singing, as she did not spit during that time.

Risk for constipation and urinary incontinence. Mrs. James could not tell staff when she needed to use the bathroom or was uncomfortable, i.e., constipation. Staff observed for nonverbal signs of discomfort, i.e. pacing and pulling on clothes, and then they escorted her to the bathroom. Bowel and bladder functions were charted and appropriate interventions implemented.

3. What aspects of the facility’s approach were central to ensuring Mrs. James care and quality of life?

Meeting with her daughter and incorporating information from the Daily Care Snapshot provided the foundation for individualized care. Staff received training in semantic PPA and knew that Mrs. James had increasing trouble with verbal cues and conversation as her understanding of words declined. Staff accommodated Mrs. James rigid and ritualistic behavior, rather than expect her to change. They built on her intact strengths and interests where possible. Playing computer solitaire adapted her executive secretary skills. Singing, favorite foods, walking, afternoon nap, and attending church, accommodated preference for set routine and interests. The facility routine was adapted to her schedule where possible, i.e., shower at 7 a.m. Mrs. James enjoyed familiar music and was encouraged to participate in musical programs. Music may be disproportionately preserved in sv-PPA compared with other modalities of knowledge.

4. What communication techniques did the staff employ that were specific to her diagnosis and adapted as her disease progressed?

Visual, verbal and physical cues were all utilized, i.e., during shower time. *Relying only on verbal cues can lead to frustration.* Visual cues were provided in context (shower and clock). Staff always greeted Mrs. James by her name, said who they were and explained why they were there to assist her. They were attentive to her verbal, “What’s a shower?” and non-verbal responses, i.e., refusals. Mrs. James was unable to read facial expressions, non-verbal responses and emotions. While it was challenging for staff to understand her emotions and responses, they did not disregard this aspect of communication. Mrs. James responded positively when a warm, easy-going, calm and quiet voice was used. Though she could not “read” emotion, she responded to the feelings conveyed by tone of voice or body language.



Managing Symptoms of Semantic PPA

Early diagnosis of semantic variant PPA (sv-PPA) is important for effective care planning. Individuals with sv-PPA lose the meaning (or semantics) of words and may develop difficulty recognizing faces and understanding the emotions of others. In addition to these cognitive changes, disinhibited, rigid and compulsive behaviors eventually develop. Some individuals retain insight and may exhibit symptoms of depression.

Tips for managing communication and behavior changes include:

- Develop speech therapy techniques to maximize communication at each stage of progression.
- Maximize information from family to learn the resident's interests, routines and key family and friends.
- A specific goal or care issue should be identified with family and staff, and speech-language interventions should be integrated into the overall care plan.
- Repeated experience or training with personally familiar objects may improve naming and single-word comprehension early in disease.
- Aided approaches (e.g., pictures, simple written words, personalized conversation boards or technology apps) that put words/pictures in front of the person help access vocabulary of daily activities.
- Monitor for eating non-food items (e.g., cat food, coffee grounds, household cleansers).
- Monitor use of shavers, knives, tools, etc. as the meaning of these items and how to use them may be lost.
- Consider nutritional drinks to supplement diet if food fads persist, or diet choices become severely limited.
- Use pictures, words and gestures in combination for context, e.g., pointing to the picture on bathroom door and word "shower" and then pointing to the actual shower was successful.
- If possible, assign same staff members on a regular basis.
- Tell the resident who you are and what you do.
- Astutely observe non-verbal behavior. Individuals may be unable to understand the meaning of bodily sensations, such as saliva. Some patients develop a preoccupation with these internal sensations.
- Evaluate all complaints with good physical examination.
- Try to incorporate resident's routine if possible.
- Posting the resident's schedule provides reminders to the staff, visitors and possibly resident.
- Focus on the present, the here-and-now rather than reminiscence therapy or projections about the future. Recent memories are much better than autobiographical memories, a pattern opposite to that in Alzheimer's disease.
- Identify and maximize retained strengths and interests to develop individual interventions build on personally meaningful history.
- Personally program the resident's iPod. There is a suggestion that music may be disproportionately preserved in sv-PPA.
- Always use a warm, easy-going, calm and quiet voice. An individual with sv-PPA may lose the ability to read emotion, but will still respond at an emotional level to non-verbals.
- Consider that visuospatial skills and calculations may remain throughout the disease. These may offer hidden strengths for creative interventions.