

Primary Progressive Aphasia, Non-Fluent Type

In order to serve people with the frontotemporal degeneration language disorders effectively in day services and long-term care, it is important to appreciate how the disease affects the patient and family over time. This case study follows one patient from diagnosis to admission to assisted living.

Early disease, October 2007:

Lilly Noble is a 46-year-old wife and mother of three teenage boys. Lilly holds a Ph.D. in English literature and is a professor at a nearby university. Over the past year, she has had some difficulty with teaching because she “loses words” and “gets stuck” in the middle of a sentence. She seems to know what she wants to say, however struggles with pronunciation. Six months ago her students reported her to the department head who insisted she be evaluated by her physician, who diagnosed the problem as early menopause. As the condition worsened she was referred to a behavioral neurologist who diagnosed primary progressive aphasia, non-fluent type (PPA), also known as “agrammatic PPA.” Last month, Lilly was placed on short-term disability and will qualify for long-term disability in six months.

Lilly’s husband Richard, a professor of engineering, advocates for Lilly by making sure she has speech and language evaluations and by teaching their sons how to best communicate. At this point Lilly is able to comprehend what is said to her, but cannot answer consistently. She is aware of the problem and expresses anxiety and frustration when pressed to speak. Her sons have learned to tell their mom what they need her to know and ask her only “yes/no” questions.

Lilly continues to keep her home, shop, and drive. She wears a MedicAlert bracelet and carries cards and a letter from her physician explaining her illness should someone believe she is having a stroke she be stopped by police for any reason. Without these, someone might charge her with driving under the influence. Adaptive devices for the telephone are employed and Lilly wears a Lifeline™ in case of emergency.

Mid-disease, October 2010:

Now 48, Lilly continues to live at home with Richard and her two sons (the eldest attends college). She is still able to say a few words, but must struggle for all but automatic speech. She understands the simpler things that are said to her, but not more complex verbal communications. She is able to point to what she needs and understands family gestures. Formerly an avid reader, Lilly now has little reading comprehension. This has made family communication difficult, and she often demonstrates fear, anxiety and anger towards family members – especially her “fast-talking” teenage sons. Her eldest son returned home to spend the summer, but Lilly did not recognize him consistently and wanted him out of the house. Lilly expresses anger by striking out, pinching, and throwing and breaking things. She has begun to kick the dog. Fearing for the dog, Richard gave it to a loving family.

Lilly is now obsessed with game shows on television, spending hours looking at them. She no longer does housework or cooking, but plays games and does searches on the computer.

Partners in FTD Care is an education initiative of the Association for Frontotemporal Degeneration that brings together health professionals, experts and families to promote understanding of FTD and to develop best practices in community care.

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Join the conversation of the Partners in FTD Care education initiative. Through the on-line forum, you can ask questions of our experts and share ideas with professionals in adult day services, long-term care, home health, hospice and other health care services who serve people with FTD. Together we will develop best practices in care planning and interventions. [Register online now!](#)

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Lilly has become hyperoral, eating huge quantities of food from the refrigerator. Richard has resorted to locking the fridge and pantry. When eating, Lilly stuffs her mouth without bothering to swallow. Fearing she will choke, Richard cuts her food in bite-sized pieces and serves very small portions every hour. Eating large quantities of food has produced periodic episodes of bowel incontinence often in public places like church and the supermarket.

Lilly has become lax in her bathing and grooming. She refuses assistance and will not wear adult incontinence briefs. Richard finally replaced her underwear from her dresser with pull-ups.

Lilly spends her day pacing indoors and attempts to elope. She is now essentially disengaged from her sons and, while insisting on being in the same room as Richard, displays no affection or gratitude. Richard and his sons are attending an FTD support group and counseling. Richard's avowed goal was to keep Lilly at home for the duration of her illness, but he is now reconsidering due to the ever-increasing burden of care and the effect on their sons.

Severe Disease, October 2012:

In February, Lilly became mute. With the exception of grunting noises or screams, she is quiet. She does not understand written or spoken language. One day in April, her teenage sons came home from school and, unable to recognize them, Lilly began to scream and cry. She was treated with an antidepressant and as time went on, an antipsychotic. The medications helped a bit but outbursts occurred with increasing frequency. By August, Lilly permanently responded to them as strangers. While she continues to "know" Richard, she no longer lets him touch her or provide care. In June, Richard hired a home health aide for bathing and respite. Finally, in September, Richard decided he could no longer manage at home. He needed to return to work in order to prepare for paying for his sons' educations and his own retirement.

Lilly was admitted to "Independence Square," an assisted living facility with a memory care unit for people with Alzheimer's disease. While the staff had previously worked with other clients/patients with language problems, they were eager to learn strategies tailored to Lilly's behaviors.

The following case plan was developed for Lilly:

- Lilly was admitted to a private room and her windows secured closed.
- Lilly has a daily routine that is followed consistently.
- Lilly does not socialize with other residents and eats alone and sits/stands at the outskirts of activities.
- Staff are trained to speak in one- or two-word phrases and watch for her non-verbal responses.
- Lilly spends much time pacing, so a circular route was established including halls and activity room. Stations were developed including putting a recliner near a window with a bird feeder (and another at an aquarium), to encourage her to sit and watch.
- She is given snacks every 30-60 minutes that are cut into bite-sized pieces.
- Lilly is now having some difficulty coordinating her tongue, lips and muscles for swallowing. Food consistencies have been modified for easier swallowing, and staff supervise all eating behaviors to prevent/alleviate choking.
- Lilly has developed a disengaged stare that some staff members, families, and other residents interpret as hostile. Staff has been trained to reassure others that the "stare" means nothing.
- Lilly will sit and color for 15-20 minutes alone in the dining area throughout the day. Special crayons are used to prevent coloring everything.

TROUBLES & TIPS

Types of PPA

Q: If non-fluent/agrammatic is one type of PPA, what are the others? How do the subtypes differ?

A: Primary progressive aphasia (PPA) is a language disorder that affects a person's ability to speak, read, write and understand what others are saying. It is a disease process that affects the frontal and temporal areas of the brain. PPA is distinct from aphasia caused by stroke. In 2011, PPA was reclassified into three clinical subtypes: nonfluent/agrammatic PPA, semantic PPA and logopenic PPA.

Nonfluent/agrammatic PPA is deterioration in the ability to produce speech. People first become hesitant in their speech, then begin to talk less and eventually become mute. Current research suggests that the fundamental loss in nonfluent/agrammatic PPA is deterioration in knowledge of the grammatical organization and the production of sounds for language.

The hallmark of **semantic PPA** is difficulty generating or recognizing familiar words. For example, when a person is shown a picture of a cat, he can neither name it nor can he recognize the word when it is provided and may ask "what is *cat*?" when it comes up. This typically affects rare words first and common nouns in later stages. Verbs and abstract words are surprisingly spared and fluent spontaneous speech is retained. Some people have problems recognizing familiar objects and faces, which can help confirm the diagnosis.

The presenting feature in **logopenic PPA** is deterioration in the ability to retrieve words. People present with a slow rate of speech with frequent pauses due to significant word-finding problems. Current research suggests that the fundamental loss in logopenic PPA is in phonologic short-term memory, which also contributes to difficulty with sentence and phrase repetition, and understanding long or complex sentences.

For more information on PPA, visit [AFTD's website](http://www.theaftd.org).



- Lilly resists care; however, using a Towel Bath™, the staff are able to keep her clean and comfortable.
- Lilly began to smear her excrement, so the staff uses jumpsuits to prevent access to the inside of her undergarments.
- Richard and his sons continue to visit and have developed strong bonds with the staff.

Questions:

1. **What symptoms does Lilly exhibit during each stage of the disease? What losses does she experience?**

Early Disease Stage (age 46)

Lilly develops communication issues. She begins to “lose words” and “get stuck” in the middle of a sentence. She struggles with pronunciation and cannot respond consistently. Lilly is aware of her difficulties and experiences anxiety and frustration when pressed to speak. Loss of language fluency and her diagnosis with PPA led to loss of a rewarding career in academia and placement on disability.

Mid-Disease Stage (age 48)

Communication challenges increase. She is only able to say a few words and does not understand complex verbal communication. Reading comprehension is now limited. As the disease spreads to other areas of the brain, Lilly experiences changes in behavior control. She responds to family members with fear, anxiety and anger. Obsessed with game shows, she no longer takes care of the house or cooks; her personal hygiene declines. Relationships are difficult to maintain. She displays no connection to her sons or affection or gratitude toward her husband.

Severe Disease Stage (age 50)

Lilly has become mute, producing just grunts or screams. She does not understand written or spoken language. Her increased need for hands-on care and resistance to the help of family prompt Richard to pursue a specialized dementia assisted living facility. The staff assesses her needs, incorporates input from family and implements an individualized plan based on a consistent, low-demand routine, a supportive structure and monitoring for safety.

2. **How do Lilly’s symptoms affect her family?**

Lilly becomes increasingly dependent on her husband and sons to provide a safe environment and direct care for her daily needs. Adjustments to the physical environment (locking the fridge and pantry) and emotional and behavioral changes increase stress on the household. Financial stress from Richard’s full-time caregiving increased and necessitated a return to work. Richard admits Lilly to an assisted living facility due her refusal of care and the increasing impact on their sons. He and the sons attend an FTD support group and counseling, and stay involved and supportive of Lilly and facility staff.

3. **How does knowing the trajectory of Lilly’s disease help the facility staff care for Lilly?**

The family provides insight into Lilly’s life (an English literature professor, mother and wife) prior to PPA and to how PPA changed Lilly’s life and the lives of her family. A tool like AFTD’s Daily Care Snapshot invites family input. It assists with her case plan to understand safety issues of elopement, swallowing difficulties and her resistance to care; appreciate communication needs/approaches; and help with her interpersonal style and lack of socialization--pacing, disengaged stare and need for individual structure. It establishes her current abilities--coloring, snacks, and non-verbal communication and initiates a strong bond between staff and family.

AFTD’s COMMITTEE ON LONG-TERM CARE EDUCATION

Partners in FTD Care is directed by a committee of professionals and caregivers dedicated to improving care for people diagnosed with FTD and their families. The AFTD Committee on Long-Term Care Education includes:

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