The Association for Frontotemporal Degeneration

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AFTD's Digital Info Packet

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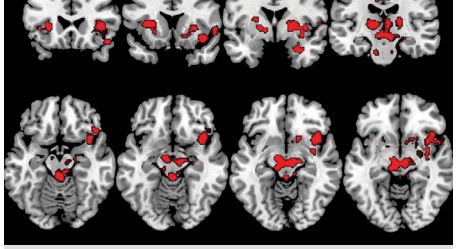

About AFTD

The Association for Frontotemporal Degeneration (AFTD) is a nonprofit founded by Helen-Ann Comstock and a host of dedicated volunteers in 2002. We work every day to advance:

Research We promote and fund research toward diagnosis, treatment and a cure.

Awareness We stimulate public awareness and understanding.

Support We inform and support those directly impacted.

Education We promote and provide education for healthcare professionals.

Advocacy We advocate for research and appropriate, affordable services.

We envision a world with compassionate care, effective support, and a future free of FTD. We hope you'll join with us in making that world a reality.

HelpLine: 866.507.7222 or info@theaftd.org

facebook.com/TheAFTD

twitter.com/AFTDHope

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We envision a world with compassionate care, effective support, and a future free of FTD.

AFTD The Association for Frontotemporal Degeneration
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FTD is the most common form of dementia for people under 60.

Short for frontotemporal degeneration, FTD represents a group of brain disorders caused by degeneration of the frontal and/or temporal lobes of the brain. It is also frequently referred to as frontotemporal dementia, frontotemporal lobar degeneration (FTLD), or Pick's disease.

How does FTD differ from Alzheimer's disease?

1) **Different symptoms.** FTD brings a gradual, progressive decline in behavior, language or movement, with memory usually relatively preserved.
2) **It typically strikes younger.** Although age of onset ranges from 21 to 80, the majority of FTD cases occur between 45 and 64. Therefore, FTD has a substantially greater impact on work, family, and the economic burden faced by families than Alzheimer's.
3) **It is less common and still far less known.** FTD's estimated U.S. prevalence is around 60,000 cases, and many in the medical community remain unfamiliar with it. FTD is frequently misdiagnosed as Alzheimer's, depression, Parkinson's disease, or a psychiatric condition. On average, it currently takes 3.6 years to get an accurate diagnosis.

How does FTD progress?

The progression of symptoms—in behavior, language, and/or movement—varies by individual, but FTD brings an inevitable decline in functioning. The length of progression varies from 2 to 20+ years.

As the disease progresses, the person affected may experience increasing difficulty in planning or organizing activities, behaving appropriately in social or work settings, communicating with others, or relating to loved ones.

Over time, FTD predisposes an individual to physical complications such as pneumonia, infection, or injury from a fall. Average life expectancy is 7 to 13 years after the start of symptoms. The most common cause of death is pneumonia.

Is treatment available?

Today, there is no cure for FTD. Unfortunately, no current treatments slow or stop the progression of the disease. However, important steps can be taken to preserve and maximize quality of life. A growing number of interventions—not limited to medication—can help with managing FTD symptoms. Support groups are a crucial source of information and connection. Our website, www.theaftd.org, offers practical information about disease management and care coordination.

AFTD funds groundbreaking research to foster timelier, more accurate diagnosis, treatments, and a cure for FTD. Today's research can lead to tomorrow's breakthroughs—but only with support from our community.

Together, we can make a difference.

AFTD is community-driven, and our work to #endFTD can move more quickly with your help—whether as a donor or a volunteer.

Our nationwide volunteer and grassroots events programs **build awareness** and raise funds.

AFTD-funded grants and awards help spur **cutting-edge research**.

With help from volunteers, **AFTD leads in advocacy** for greater federal funding for research and programs that provide affordable, high-quality care.

Learn more at **www.theaftd.org**

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SHARE HOPE.

What's Next When the Doctor Says It's FTD?

A Checklist for Navigating an FTD Diagnosis

Pause to consider your approach.	
Know that although the path to a diagnosis is often difficult, life doesn't end when FTD is diagnosed.	
Engage with people who understand FTD as soon as possible. Visit AFTD's website to find resources and support (**www.theaftd.org**).	
Contact AFTD's HelpLine with questions and to find support: call **866-507-7222** or email **info@theaftd.org**.	
Start important conversations about care and support and consider how things may change as the disease progresses.	
Identify ways to adjust and maintain the things that are most important to both the person diagnosed and close family or friends.	
Share information about the disease and about your needs with key family and friends.	
If you have children and teens, get AFTD's booklet *What About the Kids?* and visit **www.AFTDKidsandTeens.org**.	
Learn about the disease.	
Confirm the diagnosis as best you can. Consider traveling to an FTD center for a second opinion.	
Learning is ongoing: Continue to read about the disease and ask questions.	
Start a file of key articles and resources on FTD that will help you to educate others.	
Double-check information found online. Use websites you can trust and confirm with experts.	
Create your care team.	
Identify medical professionals and healthcare providers and coordinate the services they provide.	
Obtain copies of diagnostic evaluations for your records. Keep paperwork organized.	
Keep a log or journal of significant changes in symptoms. Prioritize issues to address with a doctor.	
Maintain a chronological record of all medications started and discontinued.	
Consult occupational, physical and speech therapists for evaluation and techniques to maximize abilities.	
Explore FTD-specific supports for care partners and the person with the disease. Visit **www.theaftd.org** to explore support options in your state or online.	
Keep a list of what you need. Ask family, friends and neighbors to help.	

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What's Next When the Doctor Says It's FTD?

Address legal and financial issues.	
Consult an Elder Law attorney and/or financial planner.	
Plan transition from employment, if still working.	
Complete legal documents (examples include Power of Attorney, living will, trusts, etc.).	
Review finances and make long-term plans for care.	
Apply for Social Security Disability (Compassionate Allowances Program).	
Determine eligibility for Veterans Administration benefits.	
Focus on wellness and a positive daily routine.	
Follow a regular daily routine to structure the day. Try to incorporate a heart-healthy diet and regular exercise.	
Stay active with friends and interests. Adapt activities according to strengths and needs.	
Review and visit day programs and long-term care facilities in advance of possible placement.	
Use professional counselors to help cope with changes.	
Attend an FTD education conference, either online or in-person where feasible and safe for you and your family.	
Apply for AFTD's Comstock grants—these can be for care partner respite, travel for in-person FTD conferences, or a quality of life stipend for the person with FTD.	
Address safety issues.	
Consult guidance on navigating an FTD journey during the COVID-19 pandemic on our website: **www.theaftd.org/living-with-ftd/covid-19-and-ftd/**	
Assess home environment for safety and risk regularly. Make changes as needed before a crisis occurs.	
Carry complete ID with emergency contact information. Include statement about neurological disorder and/or FTD.	
Where judgment is impaired, monitor bank accounts, investments, and online activity; change access as needed to protect assets.	
Use GPS monitoring or similar device if getting lost is a risk.	
Learn the laws where you live regarding driving privileges. This resource on our website also offers useful guidance: **www.theaftd.org/driving-and-ftd/**	
Participate in research.	
Follow emerging research to understand issues important in FTD.	
Join the FTD Disorders Registry: **www.FTDRegistry.org**	
Become familiar with observational studies, clinical trials and opportunities to participate.	
Consider autopsy and/or brain donation to confirm diagnosis and advance research. Plan early if interested in brain donation for research.	

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Criteria for Behavioral Variant FTD

In 2011, an international consortium developed revised guidelines for the diagnosis of behavioral variant frontotemporal dementia based on recent literature and collective experience. The following chart delineates the new criteria for bvFTD.

International consensus criteria for behavioural variant FTD

I. Neurodegenerative disease
The following symptom must be present to meet criteria for bvFTD
- A. Shows progressive deterioration of behaviour and/or cognition by observation or history (as provided by a knowledgeable informant).

II. Possible bvFTD
Three of the following behavioural/cognitive symptoms (A–F) must be present to meet criteria. Ascertainment requires that symptoms be persistent or recurrent, rather than single or rare events.
- A. Early* behavioural disinhibition [one of the following symptoms (A.1–A.3) must be present]:
  - A.1. Socially inappropriate behaviour
  - A.2. Loss of manners or decorum
  - A.3. Impulsive, rash or careless actions
- B. Early apathy or inertia [one of the following symptoms (B.1–B.2) must be present]:
  - B.1. Apathy
  - B.2. Inertia
- C. Early loss of sympathy or empathy [one of the following symptoms (C.1–C.2) must be present]:
  - C.1. Diminished response to other people's needs and feelings
  - C.2. Diminished social interest, interrelatedness or personal warmth
- D. Early perseverative, stereotyped or compulsive/ritualistic behaviour [one of the following symptoms (D.1–D.3) must be present]:
  - D.1. Simple repetitive movements
  - D.2. Complex, compulsive or ritualistic behaviours
  - D.3. Stereotypy of speech
- E. Hyperorality and dietary changes [one of the following symptoms (E.1–E.3) must be present]:
  - E.1. Altered food preferences
  - E.2. Binge eating, increased consumption of alcohol or cigarettes
  - E.3. Oral exploration or consumption of inedible objects
- F. Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions [all of the following symptoms (F.1–F.3) must be present]:
  - F.1. Deficits in executive tasks
  - F.2. Relative sparing of episodic memory
  - F.3. Relative sparing of visuospatial skills

III. Probable bvFTD
All of the following symptoms (A–C) must be present to meet criteria.
- A. Meets criteria for possible bvFTD
- B. Exhibits significant functional decline (by caregiver report or as evidenced by Clinical Dementia Rating Scale or Functional Activities Questionnaire scores)
- C. Imaging results consistent with bvFTD [one of the following (C.1–C.2) must be present]:
  - C.1. Frontal and/or anterior temporal atrophy on MRI or CT
  - C.2. Frontal and/or anterior temporal hypoperfusion or hypometabolism on PET or SPECT

IV. Behavioural variant FTD with definite FTLD Pathology
Criterion A and either criterion B or C must be present to meet criteria.
- A. Meets criteria for possible or probable bvFTD
- B. Histopathological evidence of FTLD on biopsy or at post-mortem
- C. Presence of a known pathogenic mutation

V. Exclusionary criteria for bvFTD
Criteria A and B must be answered negatively for any bvFTD diagnosis. Criterion C can be positive for possible bvFTD but must be negative for probable bvFTD.
- A. Pattern of deficits is better accounted for by other non-degenerative nervous system or medical disorders
- B. Behavioural disturbance is better accounted for by a psychiatric diagnosis
- C. Biomarkers strongly indicative of Alzheimer's disease or other neurodegenerative process

*As a general guideline 'early' refers to symptom presentation within the first 3 years. Table reprinted from *Brain*; permission granted to AFTD from Copyright Clearance Center.
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Partners in FTD
Care

A quarterly newsletter for
healthcare professionals and families
Winter 2021

It May Not Be Parkinson's: A Look at Corticobasal Degeneration

Introduction

Corticobasal degeneration (CBD) is a rare neurodegenerative disease that results in nerve cell loss, scarring, and shrinkage of the deeper layers in the brain's frontal and parietal lobes. (Corticobasal syndrome, meanwhile, is its most common presentation based on the pathological symptoms.) Due to their similar presentations, CBD often is initially diagnosed as Parkinson's disease. Early CBD symptoms include rigidity, slowness of movement (bradykinesia), and involuntary muscle contractions (limb dystonia) and spasms (limb myoclonus), often occurring on only one side of the body. Understanding the disease prevalence and progression can assist healthcare workers in referring persons to appropriate services, while also aiding in research. The case of David M. illustrates the difficulty in obtaining an accurate diagnosis and the importance of assembling a team to assist in diagnosis, treatment, and emotional support.

The Case of David M.

David M. is a 52-year-old senior portfolio manager for an international banking firm. Consistently praised for his business savvy and financial intelligence, David holds a PhD and multiple master's degrees, and has worked his way up the corporate ladder. Two years ago, while attending a work conference, he first experienced movement issues. An avid runner who was usually quick on his feet, David found himself tripping while climbing stairs, and even simply moving from room to room. At the time, he blamed his stumbling on a lack of sleep, but a few months later, he noticed additional abnormalities: His left hand started to tremble and stiffen when buttoning his shirts and tying his shoes, and he had trouble accepting change from the cashier when ordering his morning coffee.

A few months later, while camping with his brother Jacob, David found himself unable to get out of his sleeping bag in the morning. He felt as though he was stuck on the ground, unable to move his left leg or arm. After multiple attempts to stand, he called out to Jacob (who had previously noticed David moving awkwardly while hiking). Growing worried, and without a partner at home to confide in, David told his brother about the challenges he had been facing recently. Unsure how to proceed, Jacob suggested that David see his primary doctor, who may be able to provide insight.

Due to the issues in David's extremities, his doctor suspected Parkinson's disease and referred him to a neurologist, who had him take a balance test and a grip test, among other physical assessments. While the neurologist did not issue a formal diagnosis, he told David his symptoms fell under the umbrella of atypical Parkinsonism. He recommended the drug carbidopa-levodopa to help control his Parkinson's symptoms.

While camping with his brother Jacob, David was unable to get out of his sleeping bag in the morning–he felt as if he was stuck to the ground.

ASK AN EXPERT: "CORTICOBASAL DEGENERATION" OR "CORTICOBASAL SYNDROME"?

—Ece Bayram, MD, PhD

Corticobasal degeneration can be difficult to diagnose at the clinical setting due to heterogeneity or variability in clinical presentation and lack of reliable predictors for underlying pathology. The terms "corticobasal syndrome" (CBS) and "corticobasal degeneration" (CBD) are sometimes used interchangeably, but these terms represent different entities.

In CBD, an assessment of the underlying neuropathology will show lesions in cortical and striatal neurons and glial cells that contain abnormal tau proteins, coupled with loss of neurons in the brain's cortical regions and substantia nigra. In particular, astrocytic plaques and extensive thread-like pathology are features indicative of CBD pathology.

Corticobasal syndrome refers to the most common clinical presentation of this pathology. CBS is defined by axial akinesia, Parkinsonism, dystonia, apraxia, cortical sensory deficits, myoclonus and alien limb phenomenon. Although up to 50% of persons with CBD present with CBS, they also present with other clinical presentations, all of which are associated with other pathologies. Additionally, only 50% of persons with CBS have CBD. This clinical-pathologic diversity emphasizes the importance of distinguishing the clinical syndrome (CBS) from the pathology (CBD).

This diversity makes the clinical diagnosis challenging and underscores the need for brain donation for a definite diagnosis. The low clinical diagnostic accuracy limits the ability to confidently estimate the incidence and prevalence of CBS. It also impacts the ability to conduct reliable clinical trials in this disorder, which currently lacks an effective treatment. Continuing research efforts are necessary to improve diagnostic accuracy, which will consequently lead to better participant selection for clinical trials to develop effective treatments. ■

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Path to Diagnosis

David was unsatisfied with the doctor's diagnosis, and the medication was not helping with the rigidity or tremors in his left arm and leg. His problems performing once routine tasks with his arm, which first manifested as difficulties in collecting change from cashiers, now appeared more frequently: He began to struggle with grasping and using the television remote, kitchen utensils, and doorknobs. His left leg, meanwhile, jerked uncontrollably, seemingly moving on its own; he could not fully straighten it while standing. (He would later learn that the term for this phenomenon is "alien limb.") He also began having more trouble remembering simple math equations at work and relied increasingly on a calculator for simple addition and subtraction.

Determined to get a diagnosis that would accurately explain these progressive changes, David decided to try one more doctor—a neurologist who specializes in movement disorders. In addition to routine testing, this neurologist listened attentively as David explained his symptoms and their impact on his daily functioning. Based predominately on his symptomatology, the neurologist diagnosed David with corticobasal syndrome (CBS), a neurological condition that primarily affects movement and is associated with degeneration of the brain's frontal and parietal lobes. Specifically, the neurologist based his diagnosis on the movement issues found in David's left side (which is unique to CBS); his issues with gait, coordination and walking; and his inability to perform simple mathematic calculations.

CBS is a progressive condition, and the neurologist estimated that David had four to six years to live. Despite the grim prognosis, David was relieved that he finally had a name for his condition. But he decided to keep his diagnosis to himself as to not upset family, friends, or colleagues. He established routine follow-up appointments with the neurologist who had diagnosed him; of all the doctors he had seen, he felt he had been most attentive to his symptoms.

David and his neurologist started to put together a team of specialists. The doctor said that therapy would be beneficial to him throughout the disease. He recommended the use of speech therapists, to assist with cognition, speech, and swallowing; physical therapists, to work on gait and balance concerns and aid in the use of assistive devices; and occupational therapists, to help David perform activities of daily living, thus maintaining his independence.

David decided to keep his diagnosis to himself as to not upset family, friends, or colleagues.

LIVING ALONE WITH CBD: BALANCING AUTONOMY AND RISK

—Mary O'Hara, LCSW

While living alone in early corticobasal degeneration (CBD) may be possible, it will eventually stop being a safe option as the disease advances. Progressive impairments in walking, motor skills, thinking, language, balance, judgment, swallowing and the ability to plan and carry out activities combine to prevent persons diagnosed from adequately tending to their own health and wellbeing.

Falls are one of the most common risks to health and safety for persons with dementia living alone (Gould et al., 2010, Douglas et al., 2011). For someone living with CBD, a movement disorder that falls under the umbrella of frontotemporal degeneration (FTD), those risks are magnified. Persons living alone with early CBD may also make mistakes around medications and miss medical appointments. Because of cognitive and physical changes, they are also vulnerable to self-neglect, financial exploitation, poor nutrition, and dehydration. Impairments in reasoning and insight suggest that persons diagnosed are unable to recognize their need for help, or to arrange needed services on their own (Gould et al., 2015). Additionally, many people living along with dementia, including CBD, experience isolation and loneliness, which have further negative effects on their health (Johannessen & Möller).

Ongoing Assessments and Available Supports

Nevertheless, living alone with early CBD is possible; it requires a thorough understanding of symptoms, risks, available community services and the person's own strengths. Outside assistance is also necessary to ensure that the proper safety nets are in place and one's needs are continually assessed.

To ensure that a person can safely live alone in early CBD, family members, social service agencies, and/or a guardian must monitor the situation regularly and implement care transitions as needed. If the person diagnosed is unable to understand or implement decisions around their care and refuses assistance, the next step is often a report to Adult Protective Services or the local equivalent agency (Gould et al., 2018).

Few evidence-based programs or practices exist to support living alone with dementia. However, innovative practices such as Friendly Visitor programs, dementia village models, home care services, and police programs can offer some support. In some areas, gatekeeper programs recruit mail carriers, utility meter readers, ministers, pharmacists, and others who interact with adults in their community, and train them to identify people with dementia who are living in isolation who may need assistance. These "gatekeepers" can then refer them to a central agency for resources like medication reconciliation, home-delivered meals and home care services (Gould et al., 2015).

Increasing Support Over Time

As the saying goes, "Better to plan a month too early than a moment too late." Due to CBD's progressive nature, the person diagnosed should work with their professional and family caregivers as early as possible to establish a plan for future care. Doing this early is crucial, as it allows the person with CBD to participate in making decisions about the care options available to them when they can no longer live alone. Since FTD disorders are unpredictable, it is best to be prepared with a plan – one that balances the increasing needs of the person diagnosed with their sense of self, independence, care wishes and quality of life.

In addition to consulting with occupational, speech and physical therapy services, it may help to make a list of what activities can be safely done alone, and which ones will require support to help maximize the independence of the person diagnosed. Case managers, through a local Aging and Disability Resource Center, or private care managers are excellent resources to assess one's safety and monitor their care needs over time. Helping the person to connect with peer support from in-person or online groups for people living with dementia can decrease isolation and increase quality of life.

If the person diagnosed is unaware of their limitations but has family caregivers advocating on their behalf, it is important to help them adjust to changes in their care routines. When they do not understand why additional or new supports are needed, make sure that trusted health care providers and other family and friends are involved to ease the transitions.

Supporting someone in early FTD who lives alone requires accepting certain risks while continuously adapting and implementing more support over time, as well as knowing that eventually, they can no longer live alone safely. This decision weighs heavily on persons diagnosed and families, but health care professionals can help determine when they have reached that point. As dementia diagnoses continue to increase, we must continue to talk about how to allow persons diagnosed to live alone if they choose, while still promoting their wellbeing and prioritizing their safety.

Tools for determining safe living for someone with dementia:

- National Institute on Aging—Tips for Living Alone with Early-Stage Dementia: www.nia.nih.gov/health/tips-living-alone-early-stage-dementia
- Alzheimer's Association—Dementia Care Practice Recommendations for Professionals Working in a Home Setting: www.alz.org/national/documents/Phase_4_Home_Care_Recs.pdf

Resources for obtaining additional in-home care:

- Eldercare Locator: eldercare.acl.gov
- Aging Life Care Association: www.aginglifecare.org
- Community Resource Finder: www.communityresourcefinder.org

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Managing Symptoms at Work and at Home

Determined to work as long as possible, David returned to the office following his diagnosis and continued his business as usual. Meanwhile, he began going to therapy as recommended by his neurologist. As his disease progressed, performing tasks involving fine motor skills became more difficult, but occupational therapy provided creative ways to complete his daily tasks despite his growing impairments. Physical therapy helped him loosen his muscles, which had tightened painfully in recent months, and allowed for better fine and gross motor movement. Speech therapy strengthened his vocal cords and even slightly improved his swallowing.

But as the months passed, David's job performance worsened. Colleagues increasingly questioned his work, whether in budgets or investor portfolios, and he struggled to write financial reports and file paperwork. David grew frustrated and defensive, and refused to acknowledge to himself that these errors were a result of his diagnosis.

David frequently made use of the company car to travel to off-site meetings. After several instances where he returned the car with a new set of dents and scratches, David began to be questioned by his supervisors. A coworker who often accompanied him on meetings mentioned to his bosses that David had developed a tendency to wander into the left lane while driving, and nearly collided with oncoming traffic on multiple occasions. When his supervisors confronted him, an infuriated David refused to give up the keys, and blamed the damage to the company car on other drivers and their inability to safely navigate the road.

At home, David continued having trouble with his left hand, which made cleaning nearly impossible, as he lacked the motor control and grip strength to use a broom or put away clothes or dishes. His ability to navigate stairs got worse. Still worried that news of his diagnosis would upset his family, he hired cleaning professionals to help around the house, and began sleeping on the couch to avoid going up and down the stairs, which he knew from experience could lead to falls.

David's continued troubles with his left hand made cleaning nearly impossible, as he lacked the motor control and grip strength to sweep, fold clothes, or put away dishes.

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At his office he used the elevator in lieu of the stairs. One day, however, a colleague he was accompanying to lunch suggested they bypass the line for the elevator and walk down the three flights to the ground floor. David did not want to raise any suspicions, so he hesitantly agreed. But about halfway down the first set of stairs, David's left leg stopped cooperating, causing him to fall down the remainder of the stairwell. After determining that David could not get back to his feet, his colleague immediately called an ambulance, and his company's human resources manager called Jacob, David's emergency contact.

After the Fall

Jacob met his brother at the hospital, where David revealed his CBS diagnosis. Jacob insisted that David move in with him and his family. The brothers also agreed that it was time for David to tell his boss, who thought it best that David retire early.

Jacob was happy to support his brother; watching him struggle for so long made him feel helpless, so bringing him into his home gave him a sense of purpose. But, as Jacob would later admit to his wife, he did not know what he was getting into. David became less functional over his remaining years, losing his ability to walk without support and to move his lips and face on command, drastically decreasing his aptitude for spoken language. The two brothers, who once chatted so easily about any number of topics—sports, politics, movies—now struggled to communicate at all. Jacob began feeling less emotionally connected to his brother, which put a tremendous strain on their relationship. Attending meetings—first in person, then, as the COVID-19 pandemic took hold, virtually—of a local caregiver support group that he found through AFTD helped Jacob process his grief, as he slowly lost the brother he once knew.

Eventually, David's swallowing issues grew more acute, and he entered hospice. Jacob vowed to provide support and education for people facing CBS and FTD, signing up to co-lead his local support group. He hopes that, by connecting with other families and persons diagnosed, he can shed light on CBS to improve diagnosis and foster a sense of hope for those navigating similar paths. David arranged to have his brain donated after his death. His autopsy confirmed that he had CBS and allowed David the opportunity to contribute to CBS research, so that others living with the disease may one day have a different experience.

David became less functional over his remaining years, losing his ability to walk without support and to move his lips and face on command, drastically decreasing his aptitude for spoken language.

CORTICOBASAL DEGENERATION—KNOW THE SIGNS, KNOW THE SYMPTOMS

SYMPTOMS	POSSIBLE SIGNS
Limb apraxia	• Inability to compel a hand, arm or leg to carry out a desired motion, although the muscle strength needed to complete the action is maintained • Difficulty completing familiar purposeful activity, such as opening a door, operating the television remote, or using kitchen tools • Tripping or falling
Akinesia/bradykinesia	• Absence (akinesia) or abnormally slow (bradykinesia) movement
Rigidity	• Stiffness, resistance to movement
Dystonia	• Uncontrollable muscle contraction that causes an arm or leg to twist involuntarily or to assume an abnormal posture
Cognitive	• Alien limb phenomenon—sensation that an arm or leg is not part of the body, accompanied by inability to control movement of the limb • Acalculia—inability to carry out simple mathematical calculations, such as adding or subtracting • Visuospatial deficits—difficulty orienting in space

Discussion Questions

1. David is persistent in his pursuit of a clinical diagnosis that fit his symptoms. What does this tell us about atypical Parkinsonism, specifically CBS?

Like all FTD disorders, CBS is associated with degeneration of the brain's frontal and temporal lobes. Also affected are several regions deeper in the brain that play important roles in initiating, controlling, and coordinating movement. An atypical Parkinsonian disorder, CBS is a progressive disease that presents with some of the signs and symptoms of Parkinson's disease, but that generally does not respond well to levodopa, the most commonly prescribed Parkinson's medication. Like classic Parkinson's disease, atypical Parkinsonian disorders cause muscle stiffness, tremor, and problems with walking, balance and fine motor coordination. People with atypical Parkinsonism often have some degree of difficulty speaking or swallowing; drooling can be a problem. Cognitive and behavioral changes may occur at any point in the disease. Specific clinical diagnosis can be challenging for clinicians, particularly because CBS symptoms progress and change with time, and in their early stages may overlap with other movement disorders.

2. What role did physical therapy play in David's journey with CBS?

CBS presents with multiple gait and motor function issues, such as limb rigidity, bradykinesia, postural instability, and falls. The goal of physical therapy for people with CBS is not to restore function, but to help them function within their new limitations. For David, physical therapy built strength in his muscles, allowing him to maintain his coordination for a period of time. While the literature on rehabilitation for CBS is limited, it suggests that physical therapy plays a role in managing apraxia as well as helping to maintain balance and gait.

3. Name three forms of apraxia David experienced in this case and give an example of each.

Apraxia is the inability to perform movements that typically required some practice to learn in the first place. Manual tasks or gestures become clumsy and walking can become "frozen" for several seconds at a time. The following are three forms of apraxia David experiences during his CBS journey.

1. Limb apraxia–Inability to compel a hand, arm or leg to carry out a desired motion, although the muscle strength needed to complete the action is maintained. David frequently tripped while walking and struggled to dress himself, to use the television remote and to close his fingers around objects that were handed to him.

2. Conceptual apraxia–People with this symptom are unable to perform tasks that involve multiple subtasks. As David's symptoms progressed, he began to struggle with arithmetic and administrative tasks in his workplace.

3. Facial-oral apraxia–The inability to move one's face and lips on command. Over time, David lost his ability to swallow and speech became impaired.

FROM A CAREGIVER'S PERSPECTIVE: A GUIDE TO APRAXIA IN CBS —Janet Edmunson

Apraxia is considered a core feature of CBS. However, people living with CBS and their families often find it hard to understand. Neurological textbooks provided the best summaries for my husband and me, but even with my master's degree in health promotion, many of the neurological terms they used were foreign to me—or I had forgotten them. One of those terms was apraxia.

Apraxia happens when certain regions of the cerebral hemispheres in the brain do not work properly. The main symptom of apraxia is an inability to carry out simple movements, even though a person with apraxia has full use of their body and understands commands to move. We learned that different types of apraxia affect the body in slightly different ways:

1. **Limb-kinetic apraxia**: People with limb-kinetic apraxia are unable to use a finger, arm, or leg to make precise and coordinated movements. Although people with limb-kinetic apraxia may understand how to use an object, such as a remote control, they are unable to carry out the same movement. My husband lost the ability to clap his hands or snap his fingers.
2. **Ideomotor apraxia**: This form of apraxia refers to the inability to follow a verbal command to copy the movements of others or follow suggestions for movements.
3. **Conceptual apraxia**: Similar to ideomotor apraxia, conceptual apraxia is an inability to perform tasks that involve more than one subtask. Due to my husband's conceptual apraxia, we hired an aide to accompany him to work to assist with activities of daily living and simple administrative tasks like using the telephone.
4. **Ideational apraxia**: People with ideational apraxia are unable to plan a particular movement. They may find it hard to follow a sequence of movements, such as getting dressed or bathing.
5. **Verbal apraxia**: People with verbal or oral apraxia find it challenging to make the movements necessary for speech. They may have problems producing sounds and understanding rhythms of speech. My husband was very much the conversationalist and regularly enjoyed deep discussions with close friends, but his verbal apraxia brought these conversations to a screeching halt. When my husband lost his speech, he lost much more. I still cannot imagine the intense frustration he must have felt while unable to communicate. Eventually, he seemed to resign himself to his plight and gave up trying to express himself.
6. **Buccofacial apraxia**: Buccofacial apraxia, or facial-oral apraxia, results in an inability to make movements with the face and lips on command.
7. **Constructional apraxia**: An inability to copy, draw, or construct basic diagrams or figures is the core feature of constructional apraxia. This form of apraxia intrigued me most. On one occasion, at a neurologist visit, my husband was tasked with drawing a clock with the hands pointed to 2 o'clock. I was shocked to see the greatly distorted image that he drew.
8. **Oculomotor apraxia**: People with oculomotor apraxia have difficulty making eye movements on command. My husband's visual attention deteriorated. While his reflexes were OK, he appeared to have apraxia of the saccades (eye movements that help to reorient one's vision). The neuro-ophthalmologist said his deterioration must be in the left side of the brain, mostly affecting vision in his right eye. The doctor used the term "Balint syndrome," which explained my husband's difficulty in controlling where to look and his trouble fixating on objects.

Apraxia presented itself in many forms throughout our journey with CBS. It was one of the most difficult symptoms to adapt to, as it was ever progressing. If families on this journey learn about the many forms of apraxia early on, it can help them adapt to its many presentations. Employing strategies to maintain a good quality of life for your loved one can be implemented almost immediately. ■

What to Do About...
Corticobasal Degeneration

While often misdiagnosed as Parkinson's disease based on their similar pathological symptoms, corticobasal degeneration (CBD) is a distinct condition that can cause muscle rigidity, spasms, and contractions, often on just one side of the body. (Corticobasal syndrome, or CBS, is its most common presentation.) Its largely movement-based symptoms can make for an especially frustrating experience for persons diagnosed, who lose the ability to perform movements that once came naturally to them. Below are strategies for persons diagnosed, care partners, and health professionals to use when facing CBD.

Strategies for Persons Diagnosed and Family Care Partners

- Pay attention to signs and symptoms of a possible movement disorder or atypical Parkinsonism such as rigidity or stiffness, difficulty completing common movements or gestures, and falls.
- Notice if symptoms affect one side more than the other. A greater impact on the left side could be a sign of coritobasal syndrome (CBS).
- Monitoring the effects of prescriptions such as carbidopa-levodopa, which is commonly used in Parkinson's disease, may help to determine an accurate diagnosis.
- Visit a movement disorder specialist familiar with atypical Parkinsonism so they can thoroughly evaluate movement and establish a baseline for treatment.
- Include physical, occupational, and speech therapies to help manage CBS symptoms and improve quality of life.
- Relax the expected standard of performance of the person diagnosed, rather than rushing them or arguing that they are doing something too slowly or incorrectly. What is most important is that the person diagnosed is engaged in the activity and feeling positive.
- Consider adaptive equipment like large eating utensils, stabilization devices, bathtub seats, or adaptive clothing.
- Use affirming statements (e.g., "Take your time," "I'll wait," "Would you like help?").
- Foster adapted communication. Provide simple, one-step directions and allow enough time for the person to process what you are saying and respond accordingly.
- Monitor for depression and apathy. People with CBS experience frustration and loss over their diminishing movement and communication capabilities, and are at greater risk of apathy and depression.
- Care partners should also seek a support group to learn strategies from other caregivers and learn more about the disease. Support groups can help families and those diagnosed realize they are not alone.
- Create a support team to provide support and guidance throughout the disease process.
- Consult the Penn Memory Care website for resources about driving and dementia: pennmemorycenter.org/driving-and-dementia.
- Discuss care preferences in advanced illness with loved ones, and complete related legal and financial planning documents. Visit AFTD's website for more information: theaftd.org/living-with-ftd/legal-financial-planning.

Guidance for Medical Health Care Teams

- Know the signs and symptoms of atypical Parkinsonism to facilitate accurate diagnosis, treatment and support for those affected.
- Refer families to multi-disciplinary specialty centers with experience in CBS for a comprehensive evaluation and care planning. Encourage them to get more information by contacting the AFTD HelpLine (866-507-7222, info@theaftd.org).
- Refer families to support groups for emotional support and reassurance that they are not alone in this journey. Point them to the AFTD website: theaftd.org/living-with-ftd/aftd-support-groups.
- Encourage referrals to physical, speech, and occupational therapy to design care strategies to maintain independence in activities of daily living, assess the need for assistive devices for gait and balance, and monitor for problems with swallowing.
- Ataxias can be embarrassing for those with CBS, causing them to self-isolate and shrink away from social environments. Create an environment of acceptance to help them feel more comfortable and open to try new things.
- Encourage the person diagnosed and their care partner to learn about research, clinical trials, emerging therapies and compensatory tools.
- Encourage family participation in therapy sessions to train caregivers in how to help maximize their loved one's independence and lead sessions at home.
- Consider referring the person diagnosed for a palliative care consultation. Initiating hospice services early can ease the transition and provide support in the home.
- Watch the AFTD Educational Webinar on CBS and CBD, featuring Dr. Melissa Armstrong of the University of Florida: www.theaftd.org/webinar-corticobasal-syndrome-corticobasal-degeneration-basics-what-you-need-to-know.
- Listen to needs of the person diagnosed and their families. Many people with an FTD diagnosis develop depression; encourage both them and their close family members to consider talk therapy for support.

ADDITIONAL RESOURCES

- Visit AFTD's website for more information on CBS and CBD, including a downloadable fact sheet you can print out and present to health professionals who may be unfamiliar with the condition, as well as a link to a recent AFTD Educational Webinar on CBS and CBD: theaftd.org/what-is-ftd/corticobasal-syndrome
- Other organizations that can provide information on CBS and CBD include CurePSP and the Brain Support Network.
- Janet Edmunson, a former board chair of CurePSP and a guest contributor to this issue of Partners in FTD Care, has written a book about her and her late husband's journey with CBS/CBD, *Finding Meaning With Charles*, published in 2006.
- If you have specific questions about CBS and CBD, do not hesitate to contact the AFTD HelpLine at info@theaftd.org or 866-507-7222.

Partners in FTD
Care

A quarterly newsletter for
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When the Conversation Stops: Logopenic Variant Primary Progressive Aphasia

Introduction

Primary progressive aphasia (PPA) is a neurodegenerative disease that affects the parts of the brain responsible for speech and language, resulting in the gradual loss of the ability to speak, read, write, or understand what others are saying. Researchers divide PPA into three subtypes, including logopenic variant PPA (lvPPA), which is mainly characterized by difficulty with word-finding, resulting in frequent pauses while speaking. People with lvPPA, however, generally recall the meanings of words, unlike other types of PPA. While knowing the differences between PPA subtypes can be scientifically valuable, families living with PPA are better served receiving support and advice for the road ahead. The case of Tami W. demonstrates how ongoing engagement with knowledgeable professionals can help persons diagnosed with PPA and their care partners adapt to symptoms as they progress, as well as teach them how to cope with related challenges and identify means of support.

The Case of Tami W.

Tami W. works at a sports management firm in a major U.S. city, where she is one of its top agents. Never at a loss for words, she is known for her outgoing personality, witty banter, and unique ability to connect with the athletes she represents. But in her early 50s, she began having trouble speaking. Once praised for her knack for speaking off the cuff, even in front of large crowds, she now struggled with finding the right words, resulting in long, awkward pauses in conversations with clients. In public and especially at work, Tami felt increasingly uncomfortable and embarrassed with her language struggles.

While home for the holidays, Tami's 24-year-old daughter Devon noticed that her mother was speaking with unusual hesitance and frequent mid-sentence pauses, as if she could not find the right word to say next. Devon, a recent college graduate who lives four hours away, did not recall her mom having such difficulties when she visited last year. Devon also realized that Tami, who recently turned 52, had stopped calling her on the phone in recent months, instead choosing to hold conversations via text—were her new language difficulties to blame? When she asked her mom if she was having trouble thinking of the right words during conversation, Tami acknowledged that she had, and that she worried that her problems could affect her job.

Together, Devon and Tami did some research online, which further concerned them. They asked Tami's partner, Jessie, if she had noticed anything. Jessie was tentative at first—she knew how proud Tami was of her public speaking ability—but soon acknowledged that Tami had seemed less confident in her speech lately, and was at times unable to produce words, instead using placeholders like "thing" or "whatchamacallit" to refer to objects. Tami was not surprised to hear this; she had been worried that Jessie was aware of her recent language difficulties. All three decided that a doctor's visit was in order.

Devon and Jessie accompanied Tami to her appointment. After some routine tests, her primary care physician attributed the her language changes to job-related stress coupled with increased anxiety due to menopause. The doctor prescribed an SSRI to help Tami cope with her symptoms, and scheduled a six-month follow-up appointment. While Tami felt her symptoms were caused by more than just stress, she decided to try the medication anyway.

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ASK AN EXPERT: IS LOGOPENIC VARIANT PPA AN FTD DISORDER OR ALZHEIMER'S DISEASE?

by Emily Rogalski, PhD

One of the biggest challenges with PPA is understanding which type of neuropathology is causing a person's symptoms: a form of FTD or Alzheimer's disease (AD)? One reason for this challenge is there is no one-to-one correspondence between the symptoms one experiences and the type of pathology. A neurodegenerative disorder whose primary feature is language impairment, PPA occurs when abnormal proteins implicated in AD or FTD attack the language areas of the brain. Of the three PPA subtypes, logopenic variant (lvPPA) is most commonly —but not always—associated with AD pathology, which is characterized by the accumulation of two abnormal proteins: amyloid and misfolded tau proteins in brain cells.

Knowing the underlying neuropathology causing an individual's symptoms is important so clinicians can identify appropriate clinical drug trials and eventually, effective treatments (when they become available). Currently, knowing one's clinical PPA subtype is insufficient for determining the underlying neuropathology causing symptoms. Emerging biomarkers including cerebrospinal fluid, amyloid PET scans and tau PET scans are being developed, which can help determine underlying neuropathology in living individuals. For now, true neuropathologic diagnosis can only be confirmed by autopsy.

Each diagnosis is accompanied by two labels: one that describes the clinical symptoms (e.g., PPA, a concise way to describe the language loss one is experiencing), and one that describes the type of proteins implicated in causing the symptoms (e.g. AD). It is important to differentiate the terms "Alzheimer's dementia" (a label that describes the symptoms of progressive memory loss) and "Alzheimer's disease" (the name of the neuropathological abnormal plaques and tangles seen in the brain under the microscope after death).

Families of someone with a diagnosis of PPA during life may become confused when autopsy shows AD neuropathology. Is this a misdiagnosis? No—their loved one had the clinical symptoms of PPA (e.g. language loss), and these symptoms were caused by AD neuropathology (e.g. abnormal plaques and tangles).

(see EXPERT next page)

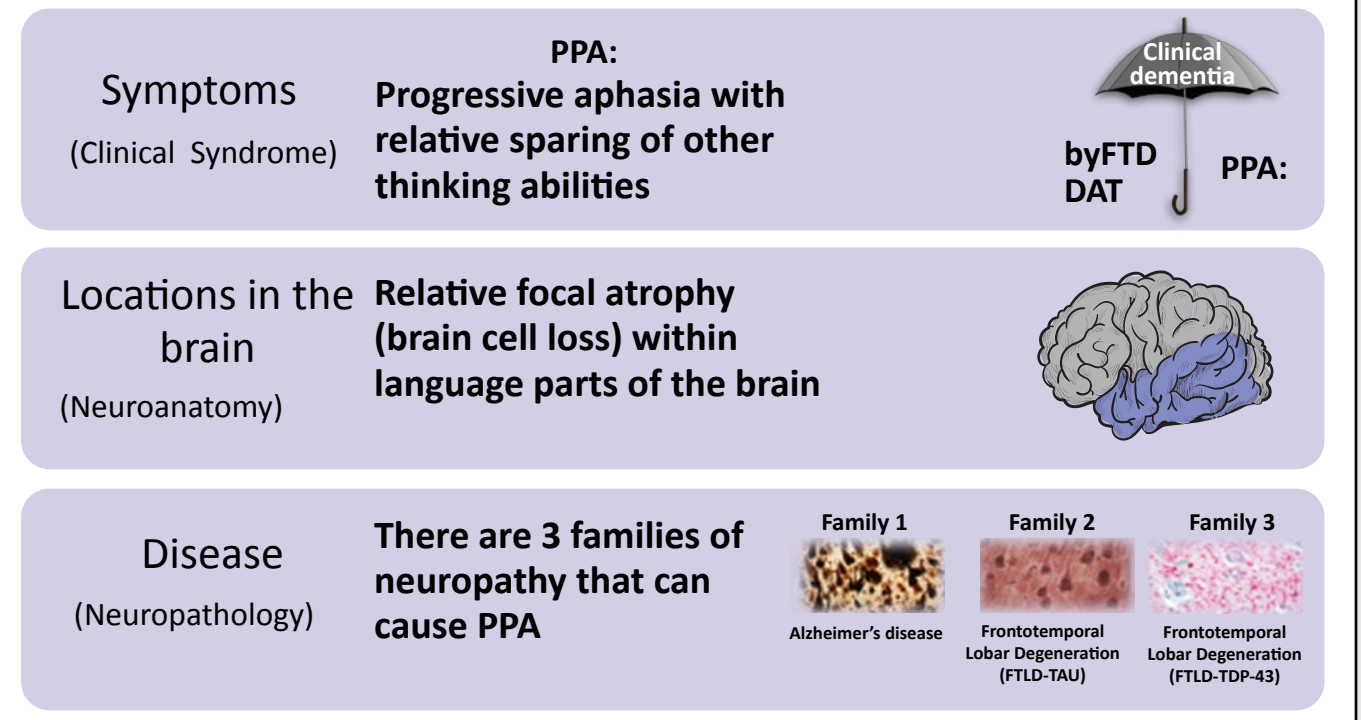
(EXPERT, continued)

Disease progression is another potential source of confusion. Neurodegenerative diseases do not stay in one location of the brain—they spread. The speed and direction of that spread is incompletely understood and variable from person to person, but is being actively studied. As the disease spreads, individuals will experience new and more severe symptoms. For some, the disease spreads to the regions of the brain that control memory; for others, the disease may spread to the frontal lobes and result in changes in personality, attention or judgment. Persons diagnosed and their families may be under the impression that PPA predominately impacts language, so the occurrence of memory loss or behavioral symptoms can cause confusion and frustration. Families can work with their clinicians, including social workers, who can help them navigate these changes.

Support groups may also be an important resource for families living with a diagnosis of PPA. Talking about PPA with others who understand the lived experience of the disease—both persons diagnosed and their loved ones—can present opportunities to learn coping strategies. While groups specifically focused on PPA can foster connections within a rare-disease community, groups focused more broadly on FTD and Alzheimer's may also help families become aware of and prepare for behavioral and cognitive symptoms that may appear over time. ■

> Knowing the underlying neuropathology causing an individual's symptoms is important so clinicians can identify appropriate clinical drug trials and eventually, effective treatments.

Fundamental Features of PPA



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Searching for Answers

Several months went by, and Tami's language difficulties did not wane. The medicine seemed to make no difference. She felt growing anxiety in her job, which is heavily language-dependent; she feared that co-workers and clients could notice her struggling to find words during meetings. Her conversational skills had long been an asset and a strength—now that talking was becoming harder, Tami experienced feelings of worthlessness, a sense of disconnect from her work, and depression.

Devon, who had started visiting more frequently, noticed her mother's condition was worsening. Her inability to remember words mid-sentence grounded conversations to a halt, and she had begun responding to questions with "yes" when she meant "no," and vice versa. Devon knew this was more than stress. Determined to find answers, she decided to seek a second and more specialized opinion. After extensive research, she contacted an institution that uses a multi-disciplinary approach to care, offering a more robust care team to provide her mother the most accurate diagnosis and personalized care.

PPA AND DEPRESSION

People with PPA experience progressive language loss, but often retain memory, personality, reasoning, and insight into their condition until the advanced stages (Mesulam, 2001; Banks SJ, Weintraub S. 2009). Engaging socially and participating in language-focused activities becomes more challenging because of their difficulties with word-finding. Combined with the awareness of their condition, those living with PPA often feel withdrawn, isolated, and excluded. Indeed, studies have shown that people with PPA are at a greater risk of experiencing depression (Medina and Weintraub, 2007).

> Attention should be given to emotional needs that are more difficult to express due to language loss.

In one study of persons diagnosed with PPA, a significant portion evaluated for depression scored in the clinically depressed range. More specifically, this study found that the number of depressive symptoms correlated with the severity of language impairments. The most common depressive symptoms included social withdrawal, lack of mental and physical energy, agitation, restlessness, sad mood and a pessimistic outlook. The study also found that people with previous depression were more vulnerable to a recurrence of symptoms because of their diagnosis (Medina and Weintraub, 2007).

Another study that compared neuropsychiatric features of PPA with cognitively normal controls suggested that PPA is associated with depression, apathy, agitation, anxiety, appetite change, and irritation. Depression symptoms are either an emotional reaction to language impairment or a noncognitive manifestation of the neurodegenerative process, the study speculated (Fatemi, 2011).

Knowing that reduced language function and a preserved understanding of their condition can leave people with PPA vulnerable for depression, the medical community must remain thoughtful and persistent in evaluating persons diagnosed for changes in mood. Attention should be given to emotional needs that are more difficult to express due to language loss. Early detection of depressive symptoms is important to ensure the most adequate treatment.

Until we have effective treatments for PPA, healthcare professionals should focus on helping those affected maintain their best quality of life. Treating depression to help improve one's mood can impact quality of life for both people with PPA and their families. Talk therapy may help, but its utility as a treatment is increasingly limited as one's PPA progresses, so non-verbal therapies such as music, art, dance and mindfulness can be considered as alternative mood interventions. Encouraging families to speak with their neurologist about their loved one's changes in mood will ensure they find the best treatment options available. ■

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At her first visit to the center, which occurred approximately two years after the onset of language difficulties, Tami underwent a series of evaluations, including a neuropsychological assessment and an evaluation with a behavioral neurologist. Imaging, blood tests and a detailed medical history were taken to help "rule in" and "rule out" different causes of her language deficits. Neuropsychological testing revealed relatively isolated deficits in language, but preservation of other thinking skills, such as memory. The neurologist found no evidence of stroke, tumor, vitamin deficiency or other potentially reversible causes of her language challenges.

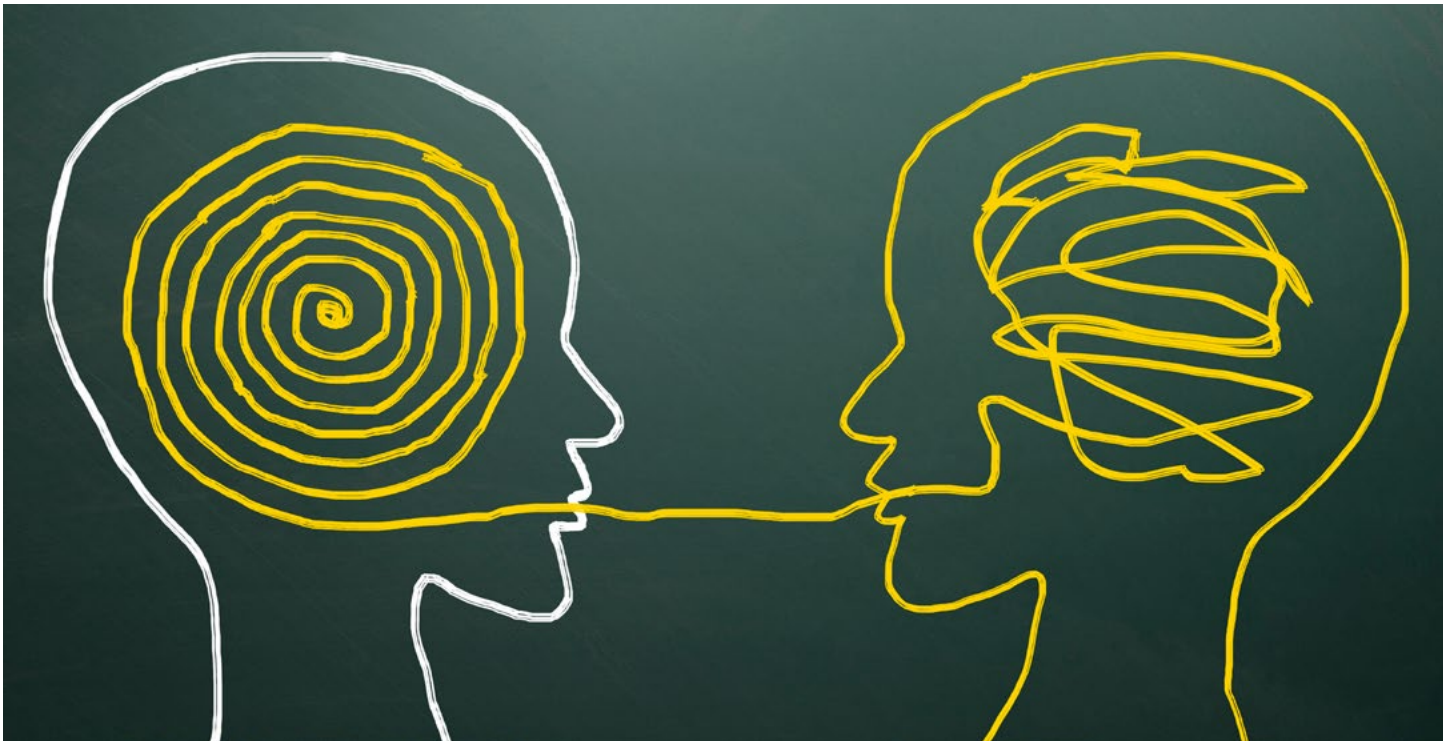
This information, in concert with the family's description of gradual, progressive loss of language, led her neurologist to make a clinical diagnosis of primary progressive aphasia (PPA). PPA is a syndrome caused by a neurodegenerative disease that currently has no cure. Tami's language challenges were most consistent with a subtype of PPA called logopenic variant PPA (lvPPA, also known as PPA-L). More specifically, people with lvPPA have difficulty recalling the names of objects and/or thinking of words in conversation, but still understand what those words mean. Persons diagnosed with any PPA subtype can expect to continue losing language skills; because the disease is progressive, it will eventually spread beyond the language areas of the brain, resulting in further changes in cognition and even behavior. Nevertheless, Tami's diagnosis gave her relief: Finally, a concrete reason for her language struggles.

Establishing a Care Plan

Tami next met with a speech-language pathologist (SLP). However, the SLP, more familiar with stroke aphasia, was uncertain how to help someone with Tami's condition, so she identified a colleague who was part of a speech and language therapy clinical trial for individuals living with PPA. The trial aimed to identify and understand which communication strategies successfully capitalize on—and compensate for—the remaining language abilities of a person with PPA. It included persons diagnosed with PPA and their care partners, recognizing that communication involves a speaker and a listener—and both would need to learn new communication techniques. After careful consideration, Tami and Jessie agreed to join the trial.

People with lvPPA have difficulty recalling the names of objects and/or thinking of words in conversation, but still understand what those words mean.

The family was also referred to a local social worker for an in-depth psychosocial assessment, which included an analysis of Tami's living situation and support networks, as well as the ways that she, Jessie and Devon were adapting to Tami's changing language abilities and overall functioning. Devon, worrying that her presence at home may become overbearing (and beginning



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to feel the pressure of putting her own career on hold to care for her mother), said she had limited her visits to every other weekend. Meanwhile, Tami and Jessie told the social worker about the strain that PPA was putting on their relationship. Jessie felt frustrated by Tami's increased dependency; for example, she had become her partner's crutch in social situations. And both grieved the loss of the way their relationship used to be. Their emotional and intimate bond was noticeably dwindling along with their ability to maintain fluid, back-and-forth conversations.

The social worker answered the questions Jessie and Devon had about PPA symptoms, and told them to visit AFTD's website for more information. She also referred them to legal counsel to secure Powers of Attorney for healthcare, finances and estate planning; and connected them to online PPA and FTD support groups to help them navigate Tami's diagnosis and treatment. Being introduced to this wealth of information helped Jessie and Devon cope. Tami, meanwhile, met with her own monthly online support group for persons diagnosed, fostering a sense of community with others living with the same rare disease.

PPA SUBTYPING: HELPING OR HINDERING THE UNDERSTANDING OF PPA?

by Emily Rogalski, PhD

One of the most confusing topics for PPA caregivers, clinicians, and researchers studying the condition has been the nomenclature associated with it. PPA subtyping—delineated as agrammatic, semantic, and logopenic subtypes—vary in clinical features and tend to have different patterns of brain atrophy, as well as different probabilistic relationships with underlying pathology. These subtypes were developed for research purposes, and were thus incompletely validated before making their way into clinical discussions.

While subtyping gives families a shorthand way to describe the constellation of symptoms a person with PPA is experiencing, there are challenges with the current diagnostic system.

On the positive side, subtyping gives families a shorthand way to describe the constellation of symptoms a person with PPA is experiencing. But admittedly, there are challenges with the current diagnostic system. Research suggests that up to 30% of individuals with PPA do not meet criteria for any of the three subtypes at the time of diagnosis. Since PPA is progressive—meaning symptoms get worse and change over time—it can also be difficult to subtype later in the disease course. And practitioners with limited specialist experience in PPA can find it difficult to diagnose the condition itself, let alone discern a subtype.

Is there an alternative to subtyping? What if a person with PPA does not meet criteria for a subtype? Neuropsychologists and speech-language pathologists with PPA expertise can provide detailed assessments of the challenges and relative strengths that the individual is experiencing. This information establishes expectations for which everyday activities may or may not be challenging for those with PPA, and can also contribute to an individualized care plan.

PPA has gotten more attention in the scientific literature in recent decades, with substantial progress being made in our understanding of the syndrome and the connections between its symptomatology, progression, pathology, and genetic implications. But many hurdles remain, particularly in terms of ensuring that people with PPA and their care partners and caregivers receive optimal information and support at diagnosis, as well as appropriate treatments, interventions and/or management strategies throughout their PPA journey.

Despite the murkiness of PPA subtyping, clinical researchers have found value in such nomenclature. But for families living with PPA, value lies not in overeducating them about their PPA subtype, but rather in anticipating their needs and equipping them with the support and information necessary to ensure the best quality of life for the road ahead. Such supports include educating them about optimizing communication to ensure the best quality of life, the role of dynamic clinical decision making in the face of declining language and other cognitive and behavioral functions, as well as emerging treatment and/or research options. ■

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Stepping Away from Work—and Toward a New Purpose

On the advice of her social worker, Tami shared her PPA diagnosis with her boss at work, who agreed that she could and should continue her work as normal. While her language ability was an issue, she had not lost the other skills—notably her ability to relate to others—that had contributed to her storied success. But over the next year, Tami noticed that exhaustion would set in each workday around noon, making it even harder to find the right words while speaking. Her boss, concerned that she may begin losing clients, eventually asked her to step down from her position and assume a part-time administrative role at the agency. While this worked out at first, Tami's language decline soon extended to reading and writing, making administrative work impossible, and she left the agency.

Tami has been able to maintain a sense of purpose in her life, allowing her to manage the grief of her diagnosis.

Tami applied for Social Security Disability Insurance (SSDI)—her social worker told her that PPA is a qualifying condition under Compassionate Allowances—but her initial application was denied. She appealed the decision, and her neurologist wrote a strong letter on her behalf, clearly stating the neurodegenerative nature of PPA and the underlying diagnosis of Alzheimer's disease or FTD. The appeal succeeded; SSDI benefits were instated and made retroactive to her first application.

Tami has been able to maintain a sense of purpose in her life, allowing her to manage the grief of her diagnosis. Since the COVID-19 pandemic took hold, she has continued to meet with the speech-language pathologist virtually, helping to improve her quality of life. Inspired by her daughter's persistence in finding an accurate diagnosis—and hoping to find meaning in the years she has left—Tami volunteered with AFTD and was eventually asked to join the AFTD Persons with FTD Advisory Council, a committee of persons living with a diagnosis who advise the staff and Board and raise awareness of the disease. She has also felt empowered through her work to advance research; after a conversation with her doctor, Tami decided to join an observational research program with eventual brain donation in hopes of helping researchers advance treatments and a cure. Through her volunteer advocacy work and participation in research, Tami has been able to not only feel a sense of personal accomplishment, but also a sense of hope in using her experience to help others in their FTD journeys.

Partners in FTD Care Advisors

The Partners in FTD Care initiative is the result of collaboration among AFTD, content experts and family caregivers. Advisors include:

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AFTD extends special thanks to all of this issue's guest contributors, including **Emily Rogalski**, PhD, and **Darby J Morhardt**, PhD, LCSW, both of the Mesulam Center for Cognitive Neurology and Alzheimer's Disease at Northwestern University, as well **Barb Murphy** and **Gary Eilrich**, both of whom contributed their perspectives as caregivers.

FROM A CAREGIVER'S PERSPECTIVE: MUSIC THERAPY AND LANGUAGE RETENTION

by Gary Eilrich

Soon after retiring, I noticed my wife was having trouble expressing herself. Our primary care physician suggested an appointment with a behavioral neurologist, and after an evaluation and neuropsychological testing, she was diagnosed with primary progressive aphasia.

She soon began attending speech therapy sessions, which she enjoyed; she was diligent about practicing her speech exercises at home. She also enjoyed and benefited from a weekly group meeting called "Speak Easy," which provided unique and innovative programing for people with neurogenic communication disorders. She attended the group most Mondays for two years; when she stopped enjoying the exercises, a speech language pathologist suggested music therapy to continue to preserve communication.

Music therapy involves an evidence-based, clinical use of musical interventions to improve clients' quality of life, and, in our case, to preserve language and communication. Always a music lover, my wife was motivated to try the therapy.

The music therapist came to our home to provide private sessions, in which I also participated so I could learn more about ways to support our communication and to know what my wife needed to practice during the week. The therapist kept meticulous notes to track my wife's progress. We learned that even though my wife was rapidly losing her ability to talk, she could still sing certain phrases, so the therapist taught her how to incorporate specific sayings into familiar songs. She learned to sing "I have to go to the bathroom," "I love you," and "The tilapia was good." (Tilapia is her favorite food.)

We spend winters in another part of the country and found another music therapist there. That therapist made a CD of phrases that she and my wife worked on so we could listen and practice on long drives. Over time, my wife's ability to express herself declined even further, but she remained able to sing the short phrases she had learned in speech therapy for longer than expected.

Music therapy provided an effective way for my wife and me to communicate even as her ability to verbally express herself declined. Most importantly, she enjoyed herself, which helped to improve her quality of life. ■

Discussion Questions

1. How did Tami's insight into her condition contribute to getting a diagnosis and her management of the disease?

Unlike in other forms of FTD, people living with PPA often retain the ability to recognize that they are changing, and can exhibit awareness or concern for the effect their symptoms have on those around them. Tami, for example, was fully aware of what was happening to her. She joined her spouse and daughter in researching her symptoms online and fully participated in doctor's appointments; later she helped to develop and facilitate her own care plan. Additionally, her self-awareness allowed her to attend and participate in a support group for persons diagnosed. The group helped Tami gain a sense of community and understanding by relating to others living with a rare, debilitating disease.

2. After Tami's diagnosis, what approach did she take to seeking help?

Tami initially met with a speech-language pathologist (SLP); however, the SLP, more familiar with stroke aphasia, was not sure how to help someone with her condition, so she identified a colleague who was part of a speech and language therapy clinical trial for people living with PPA. The intervention program included both Tami and her partner Jessie, recognizing that communication involves a speaker and a listener—and both would need to learn new communication techniques. After careful consideration, Tami and Jessie agreed to join the trial and participate in its targeted language therapies, which were adjusted as Tami's condition progressed. Throughout, they referred to the Winter 2016 issue of AFTD's Partners in FTD Care for more information about maximizing communication success with speech and language therapy in PPA.

3. What role did support groups play in Tami's PPA journey?

Support groups helped Tami and her family navigate the PPA diagnosis. Tami attended a support group specifically for people living with PPA, where she formed connections with others facing this diagnosis and learned coping skills. Jesse attended a group for caregivers, and Devon joined AFTD's Facebook group for young adults. Groups that focus more broadly on FTD and Alzheimer's provide peer support and care management strategies, and may help families become aware of and prepare for behavioral and cognitive symptoms that may appear over time.

FROM A CAREGIVER'S PERSPECTIVE: THE IMPORTANCE OF SPEECH AND LANGUAGE THERAPY IN PPA

by Barb Murphy

For people living with PPA, speech and language therapy can help slow the decline of language ability and compensate for losses over time. For my husband Gary, diagnosed with PPA five years ago, speech and language therapy has had a tremendous impact. Our decision to participate in Northwestern University's Communication Bridge Clinical Trial gave him critical word-finding and language retention skills and helped to boost his confidence when communicating.

An internet-based, speech and language therapy intervention, the Communication Bridge Clinical Trial seeks to evaluate the effectiveness of various therapies for adults with mild PPA by applying evidence-based language interventions. The 12-month program was a positive and rewarding experience, providing daily exercises to improve Gary's language abilities, as well as invaluable weekly one-on-one sessions with a trained speech-language pathologist.

Gary initially put up a fight—he said the disease was just going to worsen and nothing could change that. But eventually he agreed to participate in the program, and he's glad he did. The program's person-centered approach encourages speech-language pathologists to meet persons diagnosed at their current skill set and work towards communication goals most important to them. When we started, Gary and I were told to list words that we felt were important for him to retain, and to practice those words daily. He even practiced on vacation! Now, six months since our final session, Gary is still able to recall all but the eight most difficult words on that list.

In addition to the language exercises, I found that working with a speech-language pathologist also helped me to both better understand PPA and find ways to advance Gary's treatment. The entire team we worked with, in fact, was dedicated, patient, and encouraging. They taught my husband countless ways to search for words—skills that still help him today, almost five years since his diagnosis.

While my own participation in the program was limited—I was the cheerleader behind my husband, encouraging him when he needed it—I enjoyed seeing his dedication to practicing each day. The program's structure allowed him to track his progress, which motivated him. The skills he learned gave him the confidence to pick up the phone and call his buddies and sons for the first time since his diagnosis. For that, we are very grateful.

We have set two goals for our PPA journey: to retain for as long as possible Gary's speech and word finding—in whatever form they take; and to help others facing PPA who could benefit from this type of program. We do feel the program was helpful in our journey, and if our participation can advance research and help Gary retain those 75 words, then I am especially glad we participated. ■

To learn more about participating in the Communication Bridge Clinical Trial, please visit the Featured Studies section at theaftd.org.

PPA Resources

Publications

Khayum, B., Wieneke, C., Rogalski, E., Robinson, J., & O'Hara, M. (2012). Thinking outside the stroke: Treating primary progressive aphasia (PPA). *Perespect Gerontol, 17*(2), 37-49.

Mesulam, M.M., Rogalski, E.J., Wieneke, C., Hurley, R.S., Geula, C., Bigio, E.H., Thompson, C.K. & Weintraub, S. (2014). Primary progressive aphasia and the evolving neurology of the language network. *Nat Rev Neurol, 10*, 554-569.

Morhardt, D.J., O'Hara, M.C., Zachrich, K., Wieneke, C., & Rogalski, E.J. (2017). Development of a psycho-educational support program for individuals with primary progressive aphasia and their care-partners. *Dementia: international journal of social research and practice, 18*(4), 1310-1327.

Rogalski, E.J. & Khayum, B. (2018). A life participation approach to primary progressive aphasia intervention. *Seminars in Speech and Language, 39*(3), 284-296.

Support

FTD Phone Support: Language Group: Contact AFTD's HelpLine (866-507-7222, info@theaftd.org) for more information.

Loyola University Maryland PPA Resource & Discussion Group: www.loyola.edu/department/clinical-centers/services/specialty

Northwestern FTD/PPA Caregiver Support Group: www.brain.northwestern.edu/care-and-support/ftd-ppa-support.html

What to Do About...
Managing Logopenic Variant PPA

Introduction

The logopenic variant of primary progressive aphasia (PPA) presents a unique set of challenges for persons diagnosed, care partners and health care professionals. As language skills decline, both the person diagnosed and those around them will need to exercise patience as they develop compensatory strategies. The use of speech and language therapy techniques to cultivate remaining language strengths can be greatly beneficial. Below are some additional ways to manage the symptoms of logopenic variant PPA.

Strategies for Persons Diagnosed and Family Care Partners

- Schedule an evaluation with a specialist, such as a speech-language pathologist, familiar enough with PPA to thoroughly evaluate communication abilities and establish a baseline for treatment.
- Adapt your communication, using shorter words and phrases.
- Reduce background noise and distractions to ensure the person with PPA can understand verbal communication.
- Face the person diagnosed, speak slowly and allow time for a response. Ask for clarification if a response is incomplete or unclear; do not pretend to understand.
- Use affirming statements (e.g., "Take your time," "I'll wait," or "Would you like help?").
- Develop speech and language therapy techniques to maximize communication at each stage of progression.
- Integrate compensatory techniques (cueing, clarifying meaning, describing the word) to facilitate successful, rather than perfect, communication.
- Consider aided approaches (e.g., simple written words, personalized conversation boards or technology apps) that physically put words in front of the person diagnosed to help them access vocabulary.
- Observe triggers for behavior changes and a generalized increase in symptoms. The person diagnosed may display challenging behaviors when frustrated over their inability to communicate their wants or needs.
- Monitor for depression. People with PPA experience frustration and loss over diminishing communication and are at an increased risk.
- Explore non-verbal therapies such as music, art, dance, and mindfulness to maintain a positive outlook and to maximize quality of life.
- Participate in a support group for persons diagnosed. Groups can encourage practicing language use, as well as help to develop a sense of community with others living with a rare disease.
- Care partners should also seek a support group to learn strategies from other caregivers.
- Apply for Social Security Disability Insurance (SSDI) benefits when being a part of the workforce is no longer an option. Applying under the Compassionate Allowances program will expedite the review process.
- Consider participating in research studies investigating therapies for preserving language, such as speech and language therapy (Communication Bridge) or non-invasive brain stimulation studies (tDCS, TMS).

Guidance for Health Care Professionals

- Refer to multi-disciplinary specialty centers with experience in progressive aphasia for comprehensive evaluation and care planning.
- Encourage language evaluations and care with speech language pathologists who are knowledgeable about neurodegenerative conditions such as PPA to design strategies to retain and maximize communication abilities.
- Identify a specific goal or care issue with the family and care team; speech-language interventions should be integrated into the overall care plan.
- Listen. Tune in to what the person living with PPA and their care partners think and feel about their situation. Time spent patiently listening can create better opportunities to educate—and develop person-centered care strategies for—the person diagnosed.
- Include the spouse/partner (and, if applicable, other family members) in speech language therapy sessions to supplement at-home training.
- Pinpoint language strengths retained by the person diagnosed, and work to maximize them through individualized interventions.
- Refer to the appropriate clinician when language symptoms are coupled with behavior symptoms like depression, apathy and memory impairment.
- Encourage the person diagnosed and their care partner to learn about research, clinical trials, emerging therapies and compensatory tools.
- Reassure families and caregivers that they are not alone in their journey!
- Refer to the Winter 2016 issue of *Partners in FTD Care*, "Maximizing Communication Success in Primary Progressive Aphasia," for more information, tips and resources for managing PPA.

Behavioral Variant FTD (bvFTD)

Know the Signs … Know the Symptoms

Behavioral variant FTD, the most common form of FTD, is responsible for about half of all cases of this disease. BvFTD is also frequently referred to as frontotemporal dementia or Pick's disease.

The hallmarks of bvFTD are personality changes, apathy, and a progressive decline in socially appropriate behavior, judgment, self-control, and empathy. Unlike in Alzheimer's disease, memory is usually relatively spared in bvFTD. People with bvFTD typically do not recognize the changes in their own behavior, or exhibit awareness or concern for the effect their behavior has on the people around them. **The following are possible symptoms of bvFTD:**

SYMPTOMS	POSSIBLE SIGNS
Disinhibition A loss or lack of restraint based on social norms, leading to inappropriate behavior and impulsivity.	• Making uncharacteristic rude or offensive comments • Ignoring other people's personal space • Shoplifting, reckless spending • Touching strangers or inappropriate sexual behavior • Aggressive outbursts
Apathy Indifference or lack of interest in previously meaningful activities.	• Loss of interest in work, hobbies, and personal relationships • Neglect of personal hygiene • Loss of initiative
Emotional blunting Loss of warmth, empathy, or concern for others.	• Indifference to important events (e.g., death of a family member or friend); • Failure to recognize that loved ones are upset or unhappy
Compulsive or ritualistic behaviors Single behaviors or routines that are performed over and over.	• Repeating words or phrases • Hand rubbing, clapping • Re-reading the same book over and over again • Hoarding • Walking to the same place at the same time every day

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Behavioral Variant FTD (bvFTD)

SYMPTOMS	POSSIBLE SIGNS
Changes in Eating Habits or Diet Excessive, compulsive or inappropriate eating and drinking, or other pronounced changes in dietary preferences.	• Binge eating • Carbohydrate craving • Eating only specific foods • Increased or first-time use of tobacco products • Excessive water or alcohol consumption • Attempting to consume inedible objects
Deficits in Executive Function Poor decision-making, judgment, problem-solving, and organizational skills.	• Difficulty planning the day's activities • Questionable financial decisions • On-the-job mistakes
Other Symptoms Agitation, emotional stability.	• Pacing • Frequent and abrupt mood changes
Lack of insight As noted above, failure to recognize changes in behavior or exhibit awareness of effects of behavior on others.	• Blaming others for consequences of socially unacceptable behavior; e.g., job loss • Anger at limitations on activities

Behavioral variant FTD is *commonly misdiagnosed*—for example as depression, other psychiatric disorders, Alzheimer's disease, vascular dementia, or Parkinson's disease. If you have concern that you or a loved one may have been misdiagnosed—or about any of the signs and symptoms listed above—it is important to consult a doctor.

If you're facing this diagnosis—or if you have questions—contact AFTD for help and support.

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Corticobasal Syndrome (CBS)

Know the Signs … Know the Symptoms

Corticobasal syndrome (CBS) belongs to the category of FTD disorders that primarily affect movement. Some symptoms of both CBS and progressive supranuclear palsy, another FTD disorder associated with a decline in motor function, resemble those often seen in people with Parkinson's disease. (These features are sometimes referred to as "atypical Parkinsonism.")

Movement deficits in CBS often begin on one side of the body, but eventually both sides are affected. In addition to motor symptoms, people with CBS may exhibit changes in behavior and language skills common to the behavioral variant FTD and primary progressive aphasia forms of FTD, particularly as the disease progresses. **The following are possible symptoms of CBS:**

SYMPTOMS	POSSIBLE SIGNS
Limb apraxia	• Inability to compel a hand, arm or leg to carry out a desired motion, although the muscle strength needed to complete the action is maintained • Difficulty completing familiar purposeful activity, such as opening a door, operating the television remote, or using kitchen tools • Tripping or falling
Akinesia/bradykinesia	• Absence (akinesia) or abnormally slow (bradykinesia) movement
Rigidity	• Stiffness, resistance to movement
Dystonia	• Uncontrollable muscle contraction that causes an arm or leg to twist involuntarily or to assume an abnormal posture

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Corticobasal Syndrome (CBS)

SYMPTOMS	POSSIBLE SIGNS
Cognitive	• Alien limb phenomenon—sensation that an arm or leg is not part of the body, accompanied by inability to control movement of the limb • Acalculia—inability to carry out simple mathematical calculations, such as adding or subtracting • Visuospatial deficits—difficulty orienting in space

A person affected by CBS may present with cognitive, motor or language symptoms as the first sign. Development of a second and/or third category of symptoms makes it easier for the physician to recognize the illness as CBS.

If you have concern that you or a loved one may have been misdiagnosed—or about any of the signs and symptoms listed above—it is important to consult a doctor.

If you're facing this diagnosis—or if you have questions—contact AFTD for help and support.

Logopenic Variant PPA

Know the Signs…Know the Symptoms

FTD syndromes characterized predominantly by the gradual loss of the ability to speak, read, write or understand what others are saying fall under the category of primary progressive aphasia, or PPA. Experts further subdivide PPA into three clinical subtypes based on the specific language skills that are most affected.

People with logopenic variant PPA (lvPPA, also known as PPA-L) have difficulty finding words when they are speaking. As a result, they may speak slowly and hesitate frequently as they search for the right word.

Unlike people with the semantic variant of primary progressive aphasia, however, they are still able to recall the meanings of words. Unlike people with agrammatic PPA, speech can be perfectly fluent during small talk but then becomes hesitant and halting when the person needs to be specific or use a more unfamiliar word.

Speech is usually not effortful or distorted. The lvPPA form of primary progressive aphasia is also characterized by a narrow attention span for words that compromise the ability to repeat phrases and sentences. As the disease progresses, affected individuals may develop problems comprehending complex sentences. **The following are possible symptoms of lvPPA:**

SYMPTOMS	POSSIBLE SIGNS
Impaired single-word retrieval Difficulty finding the right word while speaking	• Pauses and hesitations due to time needed for word retrieval • Extended description (circumlocution) may be substituted for a forgotten word
Impaired repetition of phrases and sentences	• More difficulty with longer phrases and sentences
Phonological speech errors Mistakes in speech sounds, including omissions and substitutions	• Substitutes sounds made with the tip of the tongue such as “t” or “d” for sounds made near the throat such as “k” or “g”: “tup” instead of “cup” or “dap” instead of “gap” • Omits final consonants: “slee” instead of “sleep”

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Logopenic Variant PPA

SYMPTOMS	POSSIBLE SIGNS
Phonological paraphasias Substitution of a non-word with some of the same sounds for a legitimate word	• For example, the person affected may say "lelephone" for "telephone."
Poor comprehension of complex sentences	• Single-word comprehension is spared.
Difficulty swallowing	• May develop later in the disease.

Doctors will consider a diagnosis of lvPPA based on the following combination of symptoms: impaired single-word retrieval in spontaneous speech; impaired repetition of phrases and sentences; and at least three of the following: Phonological speech errors; Single-word comprehension and object knowledge unaffected; Physical ability to form words (motor speech) unaffected; and simple but correct grammar.

If you have concern that you or a loved one may have been misdiagnosed with another condition—or about any of the signs and symptoms listed above—it is important to consult a doctor.

If you're facing this diagnosis—or if you have questions—contact AFTD for help and support.

Semantic Variant PPA

Know the Signs … Know the Symptoms

FTD syndromes characterized predominantly by the gradual loss of the ability to speak, read, write or understand what others are saying fall under the category of primary progressive aphasia, or PPA. Experts further subdivide PPA into three clinical subtypes based on the specific language skills that are most affected.

The hallmark of semantic variant PPA is the progressive loss of the meanings of words. If there are additional major problems in identifying objects or faces, the condition is called semantic dementia.

Other language skills, including the ability to produce speech and to repeat phrases and sentences spoken by others, are unaffected. However, although the affected person may continue to speak fluently, their speech becomes vague and difficult to understand because many words are omitted or substituted. As the disorder progresses, people with svPPA (also known as PPA-S) may also exhibit changes in behavior similar to those seen in bvFTD, such as disinhibition and rigid food preferences.

The following are possible symptoms of svPPA:

SYMPTOMS	POSSIBLE SIGNS
Anomia Inability to recall the names of objects	• Difficulty "finding the right word" • Cannot identify a picture of a truck • Substitutes another word in the same category such as "car" for "truck"
Reduced single-word comprehension Unable to recall what words mean, especially words that are less familiar or less frequently used	• May include an inability to recognize people as well as objects • Asks "what is a truck?" when speaker uses the word in a sentence • Difficulty recognizing a family member the person does not see often
Impaired object knowledge Unable to remember what a familiar object is or how it is used	For example: • Cannot identify common kitchen utensils and how they are used in cooking

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SYMPTOMS	POSSIBLE SIGNS
Surface dyslexia/ dysgraphia Difficulty reading and writing words that do not follow pronunciation or spelling rules; such words are spelled or spoken "as if" they followed the rules	For example: • Writes "no" instead of "know" • Reads "broad" as "brode"

Doctors will consider a clinical diagnosis of svPPA when the following symptoms are observed: anomia; impaired single-word comprehension, and three of the following: Impaired object knowledge; surface dyslexia or dysgraphia; no reduction in ability to repeat words or phrases; no reduction in speech production.

If you have concern that you or a loved one may have been misdiagnosed with another condition—or about any of the signs and symptoms listed above—it is important to consult a doctor.

If you're facing this diagnosis—or if you have questions—contact AFTD for help and support.

Nonfluent/Agrammatic PPA

Know the Signs … Know the Symptoms

FTD syndromes characterized predominantly by the gradual loss of the ability to speak, read, write or understand what others are saying fall under the category of primary progressive aphasia, or PPA. Experts further subdivide PPA into three clinical subtypes based on the specific language skills that are most affected.

People with the nonfluent/agrammatic variant of PPA (nfvPPA, also known as PPA-G), also called progressive nonfluent aphasia or PNFA, find it increasingly difficult to speak yet can still recall the meanings of individual words.

The ability to form sounds with their lips and tongue is caused by degeneration of the parts of the brain that control certain related muscles; the muscles themselves, however, are unaffected. As a result, their speech becomes slow and effortful and they may appear to be physically struggling to produce words.

Speech problems alone are not sufficient for diagnosing PPA. The defining feature is the impairment of grammar. People with nfvPPA make many mistakes while speaking, including omitting small grammatical words, using word endings and verb tenses incorrectly, and/or mixing up the order of words in sentences. Eventually, some may develop difficulty swallowing as well as more widespread motor symptoms similar to those seen in the movement-predominant forms of FTD such as corticobasal syndrome. **The following are possible symptoms of nfvPPA:**

SYMPTOMS	POSSIBLE SIGNS
Agrammatism Omitting words in sentences, especially short connecting words (e.g., “to,” “from,” “the”)	• Order of words in sentences often incorrect • Errors in the use of word endings, verb tenses and pronouns • Speech restricted to short, simple phrases that are difficult for listener to understand because of omissions and errors • Uses “seed” instead of “saw” or “throwed” instead of “threw” • “Today…go lunch…ah…sister” for “today I am going to lunch with my sister.”

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Nonfluent/Agrammatic PPA

SYMPTOMS	POSSIBLE SIGNS
Apraxia of speech Difficulty producing movements of lips and tongue needed for speech	• Slow, labored speech • Groping movements of face and mouth in effort to produce correct sound • Distorted or incorrect speech sounds • Effortful speech often the first symptom
Impaired comprehension of complex sentences	• Single-word comprehension unaffected but the ability to understand long or grammatically difficult sentences is reduced
Mutism	• Affected person does not speak at all
Difficulty swallowing	• Develops later in progression of the disease
Motor symptoms	• Difficulty using an arm or leg (limb apraxia) • Slow, stiff movement • Loses balance or falls easily • Restricted up-and-down eye movement

Doctors will consider a clinical diagnosis of nonfluent variant PPA based on this combination of symptoms: apraxia of speech, agrammatism, plus at least two of the following symptoms: Impaired comprehension of complex sentence, single-word comprehension unaffected, and object knowledge unaffected.

If you have concern that you or a loved one may have been misdiagnosed with another condition—or about any of the signs and symptoms listed above—it is important to consult a doctor.

If you're facing this diagnosis—or if you have questions—contact AFTD for help and support.

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