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.

References
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). Additionally, many people living along with dementia, including CBD, experience isolation and loneliness, which have further negative effects on their health (Johannessen & Möller).

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 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.
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 sweep, fold clothes, or put away 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.
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.
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.

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:

Sandi Grow, RN, former caregiver, AFTD Board Member
Lisa Gwyther, LCSW, Duke Family Support Program
Susan Hirsch, MA, HCR ManorCare
Mary O'Hara, LCSW
Marianne Sanders, RN, University Hospitals
Jill Shapira, PhD, RN

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To join the Partners in FTD Care mailing list, or for permission to reprint this material in whole or in part, contact partnersinftdcare@theaftd.org.
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 corticobasal 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.

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**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.