Partners in FTD Care

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Falls and Dysphagia in PSP

Progressive supranuclear palsy (PSP) is a rare brain disorder that causes serious and permanent problems with control of gait and balance. Feeling unsteady when standing or walking is a hallmark symptom. People living with PSP may also experience difficulties swallowing or producing speech, and cognitive impairment. These symptoms pose distinct care challenges at home and in facilities. Because medications do not currently address PSP symptoms well, focusing on non-pharmacological interventions can impact them most effectively.

The Case of Jackie Riddle

Twenty years ago, Jackie and her husband Phil moved to a retirement community near Silicon Valley. In 2007, at age 65, Jackie began experiencing unexplained falls. A major fall late that year alerted Jackie and Phil that they were dealing with a significant issue. Within two years, she began to have trouble speaking. Her words became slurred and her speech was slow and soft, making it difficult to understand. She developed double vision.

Jackie experienced changes in thinking, including loss of executive function. For example, she was no longer able to plan and coordinate family gatherings, and she seemed unusually upset with lively dinners that featured quick-paced conversations. By 2010, she could no longer pay bills, use a computer or follow in-depth conversations. Jackie also developed a wide gait; someone said she "walked like a drunken sailor."

Despite many appointments with different neurologists and rheumatologists over four years, no diagnosis offered was able to explain these symptoms. Eventually, in 2011, Jackie saw a neuro-ophthalmologist and a movement disorder neurologist. Both specialists diagnosed her with PSP, which explained the variety of neurological symptoms affecting her gait, balance, speech, vision and thinking. While the diagnosis presented bad news, the family was also grateful to know what they were dealing with for the first time.

The movement disorder neurologist explained that most people with PSP develop dysphagia and severe swallowing problems, and that the most common cause of death in PSP relates to aspiration pneumonia or the inability to swallow. He encouraged Jackie to consider whether she wanted a feeding tube – well before the crisis of pneumonia or an inability to swallow developed. Jackie and Phil talked this over many times: Jackie was clear that she did not want a feeding tube. The couple communicated this wish to their children.

Preventing Falls

Shortly after learning the diagnosis, Phil attended a PSP support group meeting in his local area to learn about falls prevention. Falls were Jackie's first symptom and posed an immediate and ongoing risk in daily activities. Group members accepted that their loved ones' falls could not be eliminated entirely, and shared strategies to minimize falls and make them as gentle as possible when they did occur. Members recommended early consultations with physical and occupational therapists. While group members did not know of a physical therapist (PT) experienced with PSP, they recommended one with experience in Parkinson's disease, where falls are also a problem.

The PT became a valued member of Jackie's care team. She required help from in-home care aides who assisted her with getting up each morning, getting dressed, *(continued)*

Brain Changes in PSP

Progressive supranuclear palsy can be mistaken for Parkinson's disease, a much more common movement disorder. PSP is considered an FTD disorder based on our understanding of underlying biology and the location in the brain of the disease pathology. The prevalence of PSP is 6.4/100,000 people.

PSP is a tauopathy, a neurodegenerative disease associated with the abnormal accumulation of tau protein in the brain. Resultant loss of nerve cells in the midbrain region produces symptoms such as progressive postural instability leading to backwards falls (a hallmark symptom of PSP) and a relatively fixed upward gaze known as vertical supranuclear gaze palsy. Abnormal tau pathology can also appear in the frontal-temporal cortex region of the brain, leading to a prominent frontal dementia with patterns of cognitive dysfunction and atrophy similar to behavioral variant FTD (bvFTD).

Despite the similarities of PSP movement symptoms with those of Parkinson's disease, treatments that help many people with Parkinson's usually do not help those with PSP.

Sources: Lagarde et al, PLoS, 2013; Scaravilli et al, Mov Disord., 2005; Williams et al, Brain, 2007.

PSP Resources

- CurePSP www.psp.org
- "Top Resources for PSP," Brain Support Network: http://www.brainsupportnetwork.org/BSN-PSP-resources-2015-07.pdf
- "Your Personal Guide to PSP," April 2013 by The PSP Association (UK): www.pspassociation.org.uk/wp-content/uploads/2012/07/PT030-13-8-Personal-Guide-to-PSP1.pdf
- UCSF PSP/CBD Research Update, February 2014 by UCSF Memory and Aging Center: www.youtube.com/watch?v=56JAzPba59E&list=PL19WIAKy1fsN7vpgL7_Rt8iCnw18fh1j1&index=1
- LSVT-BIG physical therapy and LSVT-LOUD speech therapy for those with Parkinson's Disease. For information and a directory of clinicians see: www.lsvtglobal.com
- Parkinson's-oriented exercise videos. Find a list online at: http://parkinsons.stanford.edu/exercise_videos.html

(continued) having breakfast, exercising, bathing and getting dressed again. The PT trained Phil, Jackie and the aides how to use a gait belt to steady Jackie throughout these activities. (A gait belt is an assistive device that is secured around a person's waist to allow a caregiver to grasp the belt to help in transfers, sitting and standing and walking.) If Jackie did fall, Phil or the aide could use the gait belt to help guide her to the ground.

The PT also showed how to help Jackie get up from the floor after a fall, which was important for everyone's safety. The aides allowed Jackie to rest on the floor for as long as necessary, with a pillow and possibly a blanket for comfort. Then, when she felt able, they helped her crawl to a sofa or sturdy chair. They instructed Jackie to get up on one knee, and then, with Phil or the aide using the gait belt, provided her with gentle assistance to stand up. In this way, she was helped onto the sofa or chair to rest.

The occupational therapist (OT) evaluated Phil and Jackie's home with an eye to minimizing falls. Throw rugs were removed; a bed handle was installed to help her get in and out of bed, and grab bars were placed in the bathroom. The OT suggested the right kind of shower bench and toilet seat riser.

Like many people living with PSP, Jackie was impulsive. She would get up from a chair and start walking without her walker. Due to cognitive changes, it was not possible to educate Jackie about her own safety and have her remember to use her walker at all times. The OT suggested using a chair alarm and bed alarm so that at least someone nearby would know that Jackie was on the move.

The Importance of Exercise

The movement disorder neurologist who diagnosed Jackie told her that exercise is the only treatment that seems to slow the progression of PSP. Jackie started with a treadmill that was already in the home. Modifications were made to her exercise regimen due to the extreme risk of falls, and an "emergency off" cord on the treadmill saved Jackie from rubber burns if she fell. Later, she transitioned to a recumbent bike and did other seated exercises. The home care aides assisted Jackie with exercise as well as activities of daily living throughout her time at home.

In 2012, one year after diagnosis, Jackie had another PT evaluation and learned that it would be safer if she used a walker. The PT recommended a walker designed for people with Parkinson's disease. These walkers have larger, more durable wheels than the standard aluminum walker, *(continued)*

Troubles & Tips

Question: One of our assisted living residents has PSP. His family accepts the high risk of falls and wants him to stay active, but if someone falls after the nurse has left, we must have them evaluated at the ER. The resident has been to the ER four times this month and is sent back almost immediately. It's frustrating for both the staff and the family, and stressful for the resident. What can we do?

Answer: The high risk of falls and choking associated with PSP presents difficult challenges for facility care, and such incidents are often reasons for discharge from a residence. People with PSP and their families may understand these symptoms as consistent with the disease and be willing to accept some risks to maximize the individual's personal preferences and quality of life. Helping the family understand limits on care and what their options are can lay the foundation for success and reduce transitions.

There is considerable variability in the state, corporate and individual community regulations and policies that govern assisted living facilities. Approaches to the risk of falls and swallowing difficulties are especially important in PSP. Family may want to extend the use of a walker over full-time wheelchair use. Some facilities allow the use of chair/bed alarms to notify staff when a person stands, and these may need monitoring to prevent falls. In other facilities, bed alarms are not used as they are thought to provide a false sense of security.

The high risk of falls and choking associated with living with PSP presents difficult challenges for facility care.

Facilities often prefer to use pre-thickened liquids, which save time for licensed staff and reduce the potential for thickening agents to be mixed improperly by non-licensed staff. Pre-thickened coffee and tea are available, but some people with swallowing difficulties may find thickened liquids unappealing and unpalatable. Many assisted living facilities do not allow staff to assist with resident feeding.

In the case of Jackie Riddle, her family was told that the facility aide could put food on the utensil, and might be allowed to help raise the arm, but the aide could not help guide the utensil into the mouth. Jackie's family coordinated closely with facility administrators and had the ability to hire private aides, who thickened liquids and assisted with feeding. Some facilities use "Negotiated Risk" documents to demonstrate that families understand the risks – such as using thickener – and accept them. When there are, for example, falls or swallowing issues, staff meet with the POA, therapists and physician (if possible), and agree upon a written plan. The plan would include potential risks and possible interventions. The document is not a legal contract but assists in communicating about expectations and actions.

Hospice services can add a valuable resource for the family and the facility in the last stages of the disease. The hospice provider can carefully clarify expectations with the family and identify what interventions they are comfortable with; for example, the family may choose to waive diagnostic tests should a fall occur. Hospice can cover the cost of pain management or needed equipment. Through creative collaboration, the facility can help to avoid or delay empty beds and/or multiple transfers for residents and families.

(continued) and offer braking systems and seats so the user can rest. Phil and the home care aides continued to use the gait belt when Jackie walked with a walker. The PT also offered training on how to closely escort someone who walks with a walker.

Jackie's vision problems, including downgaze palsy and double vision, meant that walking was even more challenging because she could not see the floor. The neuro-ophthalmologist had tried prism glasses, lenses to help keep the eyes working together and aligned, to see if that would help Jackie, but they were of no benefit.

Dysphagia

Swallowing problems often begin four to six years into PSP. In 2012, Jackie began having mild dysphagia, which started with periodic coughing during and after meals. This was more of a problem at dinner, when Jackie was more fatigued. Phil and the care aides turned off the TV and eliminated other distractions during meals. Phil made sure the care aides knew how to use the Heimlich maneuver in case of choking. Jackie also began drooling, which indicated infrequent swallowing, rather than excess saliva. Also, left on her own, Jackie sometimes shoveled food, so only a certain amount could be put in front of her at any given time.

Jackie's neurologist ordered an evaluation with a speech-language pathologist (SLP). She gave Jackie exercises to do at home that would maintain strength in the swallowing muscles. Perhaps due to apathy (a symptom occasionally seen in PSP), Jackie wasn't interested in doing these exercises daily. Jackie and Phil agreed to focus on physical exercise.

Transition to Assisted Living

In 2013, Phil was diagnosed with a heart condition that caused fatigue and weakness. He became unable to assist in Jackie's care at home. At the encouragement of their children, they decided to move into an assisted living facility together. The couple lived in the same one-bedroom apartment. Phil needed no care, while Jackie needed a moderate-level of care initially, to assist her with dressing and bathing. Eventually, Jackie became bladder incontinent and needed assistance with toileting. Phil partnered with management at the facility to help them learn about PSP, develop a plan of care and arrange training in how to assist Jackie.

Assisted living facility staff were greatly concerned about Jackie's falls. Phil educated them that falls were a part of PSP and a risk that he and Jackie accepted to help her remain as active as possible. The caregivers were unfamiliar with using a gait belt or how to help someone get up from the floor after a fall. With Phil's support, the facility arranged for Jackie's PT to demonstrate these techniques during two staff meetings. Use of the new skills by the caregivers was reinforced by the administration and Phil. Staff recorded the number of falls and the severity, to report to Jackie's neurologist.

Jackie needed more rigorous leg exercise than was provided through the seated exercise class at the assisted living facility. While the facility had equipment with a recumbent seat that worked both arms and legs simultaneously, the caregivers could not consistently help Jackie with her exercise. Phil hired a fitness trainer and private-duty aides to assist with daily exercise.

When Jackie and Phil moved into the assisted living facility, Jackie was coughing with every meal. Fortunately the care staff found another couple willing to share a table with Jackie and Phil. The wife had Alzheimer's disease and was largely unaware of the noise accompanying mealtime. The facility administration ensured that the kitchen and dining staff were familiar with the Heimlich maneuver.

Jackie's neurologist ordered a modified barium swallow study, which was done by the same speech/language pathologist Jackie had seen earlier. Phil was allowed to be in the exam room and could see evidence of "silent aspiration" when liquids went down the air pipe and Jackie didn't cough. The SLP demonstrated that liquids thickened to the consistency of honey could be handled more safely. She showed that encouraging two swallows per bolus of food (a double swallow) was of great benefit. While additional strategies were tried, neither tucking the chin to the chest during the swallow (chin tuck) nor using a straw helped Jackie.

Phil and Jackie tried various liquids, pre-thickened or with thickening agents. Jackie preferred liquids thickened with SimplyThick®, especially if they were chilled. By 2014, meals were taking two hours each. Phil hired additional private-duty aides to provide assistance, as the care facility staff could not devote such time to one resident. Also, Jackie lost the ability to hold eating utensils. The assisted living staff was not permitted to provide hand-feeding, but privately-hired aides could. The administration and Phil worked together to ensure careful coordination among the entire care team.

In early 2015, seemingly overnight, Jackie lost the ability to swallow. By this time, she was largely mute. While it was impossible to have a traditional conversation with her, she knew who she was, who Phil was, and what was going on. Phil asked if she wanted to hold firm on her decision to not use a feeding tube. He asked her a series of yes or no questions, for which he asked her to hold up one finger for yes and two fingers for no. Phil learned that Jackie had not changed her mind. Hospice was contacted and provided wonderful assistance. Within about two weeks, Jackie died peacefully, surrounded by Phil and her children.

Another decision Jackie had made early on was to donate her brain upon death. Phil carried through with these wishes. Within a few months of Jackie's death, Phil received a report confirming the PSP diagnosis.

Questions for Discussion

(Use for staff in-service training or in resident-specific situations.)

The neurologist who diagnosed Jackie encouraged her to consider if she would want a feeding tube. Why?
Nearly everyone living with PSP will experience swallowing problems. The most common causes of death are related to loss of swallowing ability – either aspiration pneumonia due to severe dysphagia or total loss of the ability to swallow. Making and communicating a decision about a feeding tube and other advanced health care practices is difficult in a crisis situation. Early discussion and documentation of advanced health care preferences and power-of-attorney are especially important for any neurodegenerative disorder in which cognitive capacity is expected to diminish over time.

What strategies did the physical and occupational therapists offer to reduce Jackie's risk of falling?
The physical therapist evaluated Jackie for mobility assistance devices and recommended the gait belt and walker. The PT provided the family and caregivers specific instruction on how to escort Jackie when walking and how to help her get up off the floor safely and comfortably after a fall. Having knowledge of balance problems was important for the PT to be able to offer an exercise regimen that suited Jackie's interest and level of ability. The occupational therapist provided a safety evaluation of the home environment, with special attention to the bathroom, a room with multiple risks for people with PSP. Throw rugs were removed, a bed handle was installed and grab bars were placed in the bathroom. The OT suggested the right kind of shower bench and toilet seat riser.

What strategies did the speech-language pathologist (SLP) offer to reduce the risk of aspiration pneumonia and facilitate swallowing?
The SLP evaluated Jackie's voice, speech and ability to swallow while she was living at home. She offered Jackie exercises to maintain strength in the swallowing muscles. When Jackie transitioned to assisted living, the SLP conducted a modified barium swallow study to evaluate her ability to swallow various thicknesses of liquid, with and without a straw, with and without a chin tuck and with a double-swallow after each bite or drink. Using thickened liquids and encouraging a double swallow helped Jackie.

The Importance of Exercise

Falls are common in PSP and they are a primary symptom in about 60% of cases. Falls are often backwards and may seem to occur with no explanation: Nothing was in the path, lights were on and eyeglasses were worn, no alcohol or sedating medication had been consumed, etc. These falls can result in broken bones, which can lead to additional medical issues or even death.

Exercise is the only present treatment that seems to slow the progression of PSP, and it is very important to maintaining quality of life. Exercise helps with balance and muscle control and is the main tool in preventing falls and maximizing continued engagement in daily activities. Exercise and staying mobile help to counteract depression and mood problems, and can treat constipation and prevent urinary incontinence.

Yet the extreme risk of falls means that modifications must be made to any exercise regimen for someone with PSP. Balance exercises in particular must be modified to be made safer. If free weights are used – even lightweight ones, such as two-pound free weights – they should never be lifted above the shoulders. A physical therapist (PT) familiar with PSP can be of great assistance. Or, if the PT or a fitness trainer isn't familiar with PSP, one knowledgeable of the LSVT-BIG physical therapy program for Parkinson's is a good choice. At minimum, the PT or fitness trainer must be educated with regard to the extreme fall risk in PSP.

For all of us, metabolism slows down 90 percent after 30 minutes of sitting. But healthy individuals are constantly moving around, while PSP patients are not. A good practice is to assist the person to stand every 1-2 hours, which makes blood flow upward to the heart to be circulated throughout the body. When someone is no longer able to walk, exercise is still important. Seated exercises can keep arms and legs moving. If the person has some strength, he/she can assist with transfers such as bed to chair, and can assist with his/her own personal care longer. Due to rigidity from the disease, a person with PSP may not move at all when seated or lying down. The absence of shifting movements in bed or in a chair increases the risk of bedsores.

We extend special thanks to this issue's expert guest contributors:
Robin Riddle, CEO of the Brain Support Network and former caregiver, and
Phil Myers, Board Member for the Brain Support Network and former caregiver.

Falls and Dysphagia in PSP

Progressive supranuclear palsy (PSP) is a rare brain disorder that causes problems with control of gait and balance, and problems with swallowing known as dysphagia. Whereas medications do not currently address symptoms of balance, swallowing, or cognition in PSP, non-pharmacological interventions can impact them most effectively.

Falls

- Consult with a physical therapist (PT) experienced with PSP or Parkinson's disease for falls prevention strategies, including strategies for transferring in and out of vehicles.
- Exercise! Work with a PT or personal trainer with experience in PSP or Parkinson's to develop a modified exercise program.
- If the person walks on a treadmill, use one with an "emergency off" cord to automatically stop it in case of falls. A body-weight support treadmill can also be considered. Later, move to a recumbent bike, seated pedal exerciser and other seated exercises.
- If free weights are used - even lightweight ones such as two-pound weights - they should never be lifted above the shoulders.
- Consider the LSVT-BIG physical therapy program for Parkinson's disease, adjusted to the person with PSP.
- In consultation with a physical therapist, select a walker designed for people with Parkinson's disease for larger, more durable wheels, a braking system and a seat for the person to rest.
- Use a chair/bed alarm to alert the caregiver when the person stands and may need monitoring. Many people living with PSP are impulsive and may rise from a chair to walk without regard for safety.
- Consult with a mobility equipment specialist or PT about an appropriate wheelchair, giving consideration to preventing the wheelchair from tipping over backwards, and to special seat cushions for preventing pressure sores.
- Assist the person to stand every 1-2 hours, which makes blood flow upward to the heart to be circulated throughout the body.
- Consult with an occupational therapist for a safety evaluation of the environment and for recommendations for appropriate assistive devices. Consider adding bars, poles, or railings to make transfers easier.
- Pay particular attention to the risk of falls in the bathroom; install grab bars, a toilet seat riser and a shower bench.
- Learn how to closely escort someone when walking. Eliminate unnecessary talking when walking. If someone "freezes" when walking, learn techniques to help the person get moving again, such as: counting; clapping your hands to establish a beat; or tapping the back of the frozen leg.
- Use a gait belt secured around the person's waist or chest to steady the person through: transfers, sitting and standing; and walking.
- Recognize that the risk of falls can increase in the afternoon or evening, when persons living with PSP become more fatigued.
- After a fall, individuals should be made comfortable and allowed to rest as long as needed. Then help them to a sofa or sturdy chair, instruct them to get up on one knee, then with assistance help them to the chair to rest.

Whereas medications do not currently address symptoms of balance, swallowing, or cognition in PSP, non-pharmacological interventions can impact them most effectively.

Falls and Dysphagia in PSP (continued...)

Falls, *continued*

- Consult a neuro-ophthalmologist for evaluation of downgaze palsy or double vision, and for a possible prescription for prism glasses.
- Work with a physician to assess all medications for the likelihood that they may contribute to falls (usually due to sedation).
- If bladder incontinence is a problem, work with a neurologist to address this issue, especially for nighttime urination. It is dangerous for someone with PSP to get out of bed in the middle of the night to go to the bathroom. Consider medication, incontinence briefs, a catheter (including a condom catheter for men), a plastic urinal, a bedside commode, bladder training and lifestyle changes (no liquids after 6pm, no caffeine or alcohol in the evening).

Dysphagia

- Early in the disease's progression, help the person living with PSP to consider and make known their preferences about having a feeding tube.
- Reduce distraction (e.g. turn off the TV, create a quiet atmosphere for meals) to facilitate concentration needed for swallowing.
- Consult a speech-language pathologist for evaluation of swallowing difficulties, recommendation of exercises to maintain strength in swallowing muscles and other strategies to reduce choking.
- Use strategies to facilitate swallowing, such as determining the optimal thicknesses of liquids, keeping the chin tucked down when swallowing, and using a double-swallow after each bite or drink.
- Work with an occupational therapist or speech therapist to evaluate mealtime equipment, such as: thick-handled, weighted eating utensils; special cups; scoop plates; non-skid bowls; or plate elevators.
- Ensure that family and professional caregivers know how to use the Heimlich maneuver in case of choking.
- Present only a limited amount of food at any given time to limit the shoveling of food due to cognitive impairment.
- Use pre-thickened liquids or thicken with an agent such as Thick-It®, which is cornstarch-based, or SimplyThick®, a gum-based thickener, so that liquids can be consumed more safely. Some may prefer the taste of such liquids when chilled.
- Eliminate dry meats or cereal and things with mixed consistency (such as soup) from meals. Choose moist and tender foods, such as dark meat chicken, fish, casseroles or pastas.
- Use a blender to puree food, which may be necessary for severe dysphagia. Thicken pureed food with commercial thickeners or other products, such as oatmeal, bananas or potato flakes.
- During mealtimes, give verbal cues (such as *go ahead and swallow*, *eat slow*, *one bite at a time*, *small sips*).
- Learn warning signs of swallowing problems and possible aspiration, such as: coughing during meals; drooling; dysarthria (difficulty articulating sounds); mouth stuffing; holding food in one's mouth for 20 seconds before swallowing; choking (perhaps on saliva) not during meals; or pneumonia.
- Continue good oral hygiene as this can reduce the chances of aspiration pneumonia.
- Consider smaller, more frequent meals in an effort to keep meal times less than 60 minutes. Otherwise, it may be extremely fatiguing for the person with PSP to eat.