

Frontotemporal Lobar Degeneration (FTLD)

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PREFACE: *This session is one of three companion sessions on Frontotemporal Lobar Degeneration available on the web site www.AlzOnline.net. This session is Frontotemporal Lobar Degeneration (FTLD) and details how people may act and function as a result of changes in the frontal lobes and temporal lobes of the brain; it covers frontotemporal dementia, primary progressive aphasia, and semantic dementia.*

The second session on Frontotemporal Lobar Degeneration (FTLD) with Movement Difficulties, elsewhere on the web site www.AlzOnline.net, covers progressive supranuclear palsy, cortical basal degeneration, and frontotemporal dementia with motor neuron disease. The third session on Frontotemporal Lobar Degeneration (FTLD): Brief Discussion of Tau and Ubiquitin Proteins on the web site www.AlzOnline.net contains a brief discussion of tau and ubiquitin proteins.

The following material makes up the first session, Frontotemporal Lobar Degeneration (FTLD).

Frontotemporal Lobar Degeneration (FTLD): Introduction

Frontotemporal lobar degeneration refers to the group of progressive dementias that result from decline in brain cells which control behavior, thinking and communication¹.

[NOTE: "Dementia" is a general term that refers to decline in thinking and/or memory function while an individual is awake and alert; the decline is enough to interfere with normal daily functioning, for example on the job or at home. "Progressive" dementia progresses, becomes worse over time.]

The changes of frontotemporal lobar degeneration result from brain cell loss in the frontal lobes (the front top part of the brain) and/or the temporal lobes (both the lower left side and the lower right side of the brain). Some forms of frontotemporal lobar degeneration show early signs of movement difficulties, one group of which forms parkinsonism signs (different from Parkinson disease). As discussed above in the Preface, these conditions are discussed in the separate www.AlzOnline.net session called Frontotemporal Lobar Degeneration with Movement Difficulties.

Formerly, the term Pick's Complex grouped several frontal lobe-temporal lobe dementias such as frontal dementia, primary progressive aphasia, semantic dementia, cortical basal ganglionic degeneration, frontal-temporal dementia with motor neuron disease, and dementia with amyotrophic lateral sclerosis; the grouping included Pick's disease as a specific type^{2,3}. There are different ways of grouping these specific conditions. In this session the term **frontotemporal lobar degeneration** (FTLD) is used; it covers frontal-temporal dementia, Pick's disease, primary progressive aphasia and semantic dementia⁴⁻⁷. As mentioned in the previous paragraph the lobar degeneration with movement difficulties are discussed in the separate *www.AlzOnline.net* session called Frontotemporal Lobar Degeneration with Movement Difficulties.

Some research indicates that FTLD occur in 15 to 20% of all cases of progressive dementia^{8,9}. Alzheimer's disease is considered to be the most common progressive dementia; either vascular dementia or Lewy body dementia appears to be the second most common; and frontotemporal lobar degeneration (FTLD) now ranks third^{4,7,10}. About as many women as men have FTLD¹¹ which seems to progress over the same amount of time as Alzheimer's disease though motor neuron involvement shortens the duration of the disease.

An autosomal dominant gene may explain why researchers estimate that a heredity component may be at play in 20 – 40 % of the FTLD cases while recessive heredity appears to be rare (genes on chromosomes 17 and 3 seem to be factors)¹²⁻¹⁵; the rest of the cases seem to be sporadic^{7,11,12,16}.

Some forms of FTLD seem to relate to abnormal changes of the tau protein while some forms of FTLD have abnormal ubiquitin protein^{11,16}. Other cases may have various kinds of cell inclusions while still other cases have no distinctive abnormal cells.¹¹⁻¹³

[*Note: A tauopathy, abnormal forms of the tau protein found in the plaque of brain tissue, may contribute to people developing certain progressive dementias such as Pick's disease or frontal-temporal dementia). Alzheimer's disease is considered to be a tauopathy.*^{12,13} There is more discussion of abnormal tau protein and tauopathy in the companion session, **Frontotemporal Lobar Degeneration (FTLD): Brief Discussion of Tau and Ubiquitin Proteins.**]

Frontotemporal Lobar Degeneration (FTLD) and Alzheimer's Disease Differences
Though both are progressive dementias and both are considered to be tauopathies, major differences separate frontotemporal lobar degeneration (FTLD) from Alzheimer's disease¹⁷. FTLD usually shows up in younger adults between 40 to 60 years of age; however, symptoms have been reported to begin in people as young as 35 and as old as 75 years of age^{7,18}. On the other hand, evidence of Alzheimer's disease seldom appears before the age of 65. The onset and progress of FTLD are slow. Behavioral function and personality changes, especially social behaviors, personal conduct with appropriate timing and limits, and personal hygiene begin to decline earlier in FTLD than in Alzheimer's disease. Typically, the decline in short-term memory occurs much later in the course of FTLD while in Alzheimer's disease, the short-term memory problem

usually appears first. Language problems typically occur early in Alzheimer's disease and in some specific types of FTLD, such as primary progressive aphasia and semantic dementia. Usually language skills are maintained in frontal-temporal dementia until the decline is severe. Visuospatial abilities are typically preserved during the course of the FTLD. The APOE (apolipoprotein E4) factor that seems to be associated with Alzheimer's disease does not seem to be associated with FTLD^{19,20}.

Depression

Early in the course of FTLD or much later there may be signs of disinterest and apathy that do not reflect feeling sad. This change may be due to brain cell changes and not depression, especially when the person has little insight or self-awareness of the changes in ability²¹. However, there may be evidence of depression, such as increased irritability, poor concentration, lack of attention during interactions, sadness or a negative mood, poor appetite or sleep, or the opposite over-eating and sleeping too much^{17,22}. Withdrawal from normal activities is another sign of depression. Depression should be treated for as long as the depression lasts.

If night time sleep, day time sleep and "cat naps" add up to 14 or more hours of sleep over a 24 hour period, the person is sleeping too much. Too much sleep is not healthy and leads to problems such as infections and weak muscles.

Types of Frontotemporal Lobar Degeneration (FTLD)

The disorders that comprise frontotemporal lobar degeneration (without early onset of movement difficulties) fall into two general categories based on the clinical picture of beginning symptoms²³. The first category is behavioral and personality changes. The second category is language or communication changes. The next section describes the four types of progressive dementias that fall into the two categories.

Group 1: Behavioral and Personality Changes

- a. frontal-temporal dementia
- b. Pick's disease

Group 2: Language or Communication Changes

- a. primary progressive aphasia
- b. semantic dementia

Group 1: Behavioral and Personality Changes

Early changes that impact personality and behavior without changes in short-term memory or movement ability make up two types of progressive dementia, **frontal-temporal dementia** and **Pick's disease**, discussed below.

Frontal-temporal Dementia

In frontal-temporal dementia early changes impact personality and behavior. As family members start noticing the changes, there may be evidence of mild problems recalling specific words, especially when speaking with a group of people or answering questions after giving a prepared speech. During these early changes, typically short-term memory is unchanged. In fact short-term memory, visual-spatial ability and math skills stay strong until much later as the decline involves more and more brain cells. The decline may lead to behavior that appears self-centered and uncaring.^{1,7}

As the frontal-temporal dementia becomes more obvious, behaviors may range from disinterest and withdrawal from people and activities to the opposite, more spontaneous, inappropriate behaviors. Examples of more spontaneous behavior include inappropriate friendliness such as speaking candidly and revealing personal information to strangers or becoming angry during routine tasks on the job or at home. The inappropriate behaviors may ignore social etiquette and boundaries, such as getting too close when speaking to others and unwelcome, sexual behaviors (the tendency to touch, hug, or talk to others in inappropriate, intimate ways). Often the new behaviors are very different from the previous style of behavior for that person.

The person with frontal-temporal dementia may develop unusual eating habits, such as eating too much of a specific food. Sometimes an increased appetite, especially the craving for sweets, leads to a large weight gain over a short period of time such as a gain of 40 pounds over 6 months. The person may keep mouthing objects such as combs, pens, etc. and constantly suck or chew on them.

As the disease progresses, the person may start hoarding, for example, old newspapers and magazines, empty cans or jars that held food, broken dishes, or ragged clothing. The ability to organize an activity, to plan ahead, and to consider possibilities and consequences when making choices undergoes decline. The person may become stubborn, refusing to attend a lunch that, earlier, appeared so desirable. The opposite may occur, being rude by refusing to leave a gathering when it is time to depart. Utilization behavior, touching and using any objects within reach may become a problem, for example, picking up and eating fruit in the supermarket before paying the cashier or picking up and immediately using tools in a store. The person may pick up someone else's eyeglasses and repeatedly put them on, off, and on again. Inappropriate judgment and decisions may not consider beforehand the consequences of such steps. There is little insight into the appropriateness of the behavior or the discomfort experienced by others.

The person may become easily distracted and struggle to keep focused on a task or on a conversation. Often the lack of insight involves denying the family's descriptions of the embarrassing behaviors. The person with the progressive dementia may show surprise at the remarks of the family members. The person may become paranoid and angrily claim that the remarks are untrue. This denial happens often during the medical evaluation when the family describes changes in behavior and decline in function to the physician.

There may be changes in emotions. The person undergoing the changes may show different emotions from the feelings expressed by others in the same setting. The person may appear to be indifferent and cold-feeling or appear unusually happy, even laughing or joking inappropriately when others are angry or sad. The person may seem quiet and passive, then switch suddenly to irritable, restless, angry outbursts.

Over time, the individual may have trouble with abstract thinking and may communicate more easily with concrete phrases, names of objects or specific action steps to be taken next. With the changes of the progressive dementia may come a decline in personal grooming and personal hygiene.

Pick's Disease

Pick's disease occurs in middle age (40 – 60 years of age), typically in more men than women. Early on decline is evident in personality, social skills, judgment, decision-making, and executive function; some mild short-term memory difficulty may appear early on. The personality changes may occur many years before there is evidence of other problems.^{3,24}

Early in the course of the disease, the person may overindulge in eating, especially sweets, or eat only certain foods. The person may have an increase of sexual behaviors. Other excesses may develop, such as consuming excessive amounts of certain liquids, perhaps the same brand and flavor of a soft drink or alcohol. Repeated behaviors occur, for example, repeated hand rubbing, wearing the same clothes every day, re-reading the same news story or book several times, or walking to and from the mailbox many times each day.²⁵

As the decline continues, personal hygiene and dressing may become neglected. Some people become restless and impulsive over time while others seem to lose interest and motivation and become withdrawn. After a while, memory and language changes may occur. Often the person has limited self-awareness of the changes and little concern for the effect of such behaviors on family, friends or co-workers.

As the brain cell loss continues, indifference may seem to grow. The person may show less caring, empathy and sympathy toward others even when others express great distress or sadness. Moods may change suddenly for no obvious reason.

Movement difficulties may occur over time; the movement problems may include parkinsonism which shows up as slow movement of the body, stiff muscles, decreased display of emotion on the face, balance difficulties and difficulty walking.²⁶

When a pathology examination of the brain tissue identifies a large number of Pick's bodies (or Pick's cells) in the frontal and temporal lobes, then the dementia is called Pick's disease. Pick's disease is considered a tauopathy (abnormal forms of the tau protein, found in the plaque of brain tissue).^{5,12,13,27}

Some physicians prefer to identify the condition of a person who comes for a medical evaluation as a frontal-temporal dementia until an autopsy evaluation points to Pick's disease. The disease was described first by Arnold Pick (1892), however, Alois Alzheimer, the neuropathologist who in 1911 first reported seeing the special cells, is the one who labeled the cells as Pick's bodies. Pick's bodies are found in only 25% of the patients who have a frontal-temporal dementia.^{5,27}

Group 2: Language or Communication Changes

Communication problems result when the brain cells in the temporal lobes undergo decline. Sometimes there is minimal decline in the frontal lobes; sometimes the decline involving the frontal lobes becomes more widespread over time. The two conditions discussed in this section are first, a *nonfluent* aphasia called **primary progressive aphasia** and second, a *fluent aphasia* called **semantic dementia**.

[*Note:* Aphasia is the term given to conditions that result from decline in the language centers of the brain. The language area in the rear, top part of the temporal lobe of the brain (usually on the left side of the brain for most people) holds language information; language is expressed by its connections to the frontal lobe.]

Primary Progressive Aphasia

Primary progressive aphasia, difficulty expressing words, appears typically in middle age adults. It usually comes on slowly. The person may have difficulty finding the right word, may use the correct word but apply the wrong letters when saying (or writing) the word). They may use incorrect grammar. Despite great effort, they may be very slow in expressing the word or sentence; for this reason primary progressive aphasia is often called *nonfluent aphasia*.^{2,6,28-30}

Often, early on, people with nonfluent aphasia have good short-term memory, know what they want to say and can point to the correct phrase or sentence written on a piece of paper. As the condition progresses, they typically lose the ability to recall or recognize the written word, but can point to the correct picture. Then eventually that pointing ability is lost. They may become dysarthric (weak and clumsy in moving parts of the mouth and jaw to form the words). They may develop difficulty swallowing. Eventually they may become mute.

Difficulty with language may be the only problem; all other movement and thinking skills may remain strong till very late in the disease. Or, as the disease progresses over time, other thinking skills, and memory may be affected.

In primary progressive aphasia, an early MRI or CT shows brain atrophy in the rear language storage area of the brain (typically the left posterior, superior temporal lobe) and the front temple side of the brain (typically the left frontal lobe known as Broca's area).

As the communication ability worsens, frustration and depression may build, both in the care receiver and care provider. Working harder than normal to communicate leads to increased stress and fatigue, which may lead to further break down in communication³¹.

Semantic Dementia

Semantic dementia^{6,32} presents as difficulties in: naming, recognizing familiar words, and understanding the speech of others. There is gradual loss of the meaning of nouns and objects^{21,29}. Sometimes semantic dementia is called *fluent aphasia*, because speech, though hard for listeners to understand, usually occurs at a normal rate and seems to flow spontaneously.

Early on, the occasional use of wrong words or the occasional problem understanding the message of others indicates the beginnings of a problem. Sometimes the person uses many words to explain the one word that is hard to recall. Typically people with semantic dementia can read and write accurately. When they speak, they use the wrong words and, later, though their words are pronounced correctly, the speech does not make sense. Over time they may lose their ability to express speech and develop other thinking problems (because of additional decline in the frontal lobes) such as losing the ability to make decisions.

People with semantic dementia continue to express emotions and interest in maintaining relationships (except for the difficulties in speaking to others). They usually do not repeat a behavior over and over numerous times inappropriately, such as checking 12 or more times to see if the front door is locked or if the stove is turned off (note that checking one or twice is considered normal behavior). Since semantic dementia is a tauopathy, movement difficulties such as when walking or picking up a book from a shelf, are usually not a problem. Typically, the short-term memory is preserved until late in the course of the progressive semantic dementia.³³

Medical Evaluation

The medical evaluation of symptoms of changes in thinking, memory and movement abilities should be thorough. The medical evaluation should include a physical exam, a neurological exam, a neuropsychological exam, information about the person's medical and surgical history, the family medical history, a social history (information about education completed, jobs, skills, relationships, hobbies, interests, etc.), a list of prescribed and over-the-counter medicines as well as vitamins, minerals, herbs, etc. Blood tests should evaluate electrolytes, sedimentation rates, cholesterol, triglycerides, urea, liver functions, vitamins such as folate and B12, body hormones such as a thyroid screen, and infections. Tests of urine are also important. If there is a suspicion of seizures, an EEG (electro-encephalogram) is appropriate. When the person reports the surfacing of symptoms over a short period of time such as a few days or weeks, a spinal tap helps to rule out infection as the cause of the progressive dementia.^{24,34,35}

The medical evaluation should include an MRI (magnetic resonance imaging) or CT (computed tomography). MRI or CT films of the brain tissue that show a loss of brain cells, called atrophy, in the frontal lobes (front top part of the brain) and/or the temporal lobes (the sides of the brain) indicate frontal dementia, temporal dementia, frontal-temporal dementia or frontotemporal lobar degeneration. When just a small area of the frontal lobe or the temporal lobe shows atrophy, the person may have a more focused dementia such as semantic dementia without any other deficits. An MRI or CT that shows a larger area of the atrophy may indicate the involvement of many brain systems.^{24,34,35}

The findings on the MRI indicating a frontotemporal lobar degeneration are different from the MRI or CT pointing to a diagnosis of probable Alzheimer's disease, which show a more general atrophy all over the "cap of the brain" (the cortex or top area of the brain). Early on in the course of Alzheimer's disease atrophy usually occurs in the temporal and parietal lobes, then later it typically includes areas in the frontal lobe, and much later may impact areas of the occipital lobes.

Care Management

Living a healthy lifestyle (regular physical exercise, social activities, and brain exercise) is important for the care receiver and the family (as well as other) caregivers. Good nutrition should involve including fresh fruits and vegetables in the daily meals and avoiding lots of sweets, fats and carbohydrates, especially important for people with strong cravings for sweets. It is critical for people (both the person with the diagnosis and the caregivers) to partner with other family members, health and social service providers, neighbors, and resource people available in the community to deal over the long-term with the changes of frontotemporal lobar degeneration.

Treatment with Medicine

The physician will recommend that the person with the diagnosis stop all medicines, such as antihistamines (many anti-allergy medicines, anti-spasm medicines, sleeping pills, and cough medicines), that interfere with memory, communication, and other cognitive (thinking) functions.

Research suggests that lower levels of serotonin, a chemical in the front (frontal lobes) "cap" (cortex) of the brain important for carrying information from one neuron to the next, may be helped by SSRIs (selective serotonergic reuptake inhibitors). SSRIs have helped some patients with behavioral difficulties^{36,37}, not only depression, but also to help reduce food cravings loss of impulse control, and compulsions.^{7,37}

The brain cell changes of frontotemporal lobar degeneration (FTLD) do not primarily affect the neurons that produce acetylcholine (a chemical in the brain important for carrying information from one neuron to the next) until the later or end stage of decline^{18,36,38-41}. Therefore, according to research, the three anticholinesterase medicines which boost memory and thinking functions may not be particularly useful: 1) donepezil

(aricept), 2) rivastigmine (exelon), and 3) galantamine (razadyne, previously known as reminyl).³⁵ At the same time some research suggests that these three medicines may help aphasia (primary progressive aphasia and semantic aphasia³⁶⁻³⁹).

Whenever there is a question about prescribed medicines or over-the-counter medicines, herbs, or vitamins, etc. people should check with experts regarding the impact on health. Good sources to check regarding any therapeutic or, the opposite, negative effects of medicines, etc. are a physician **and** a pharmacist (**ask both people!**).

Care Planning

Family and paid caregivers should try to keep the schedule at home as routine, simple, safe, and pleasant as possible. Since FTLN affects judgment, limits the consideration of consequences when making decisions, and decreases sensitivity about appropriate limits, it is important to partner with or restrict some of the activities the person normally handled. The caregiver should gradually decrease responsibilities that may involve that person paying bills, handling complicated machinery, caring for others especially children, being left alone to do complicated, multi-step tasks, and driving any type of motor vehicle.^{35,42}

Driving

With FTLN there is an increased safety risk when driving any type of motor vehicles including cars, trucks, vans, motorbikes, rider lawn mowers, motor boats, etc. Changes in behavior, personality, judgment, decision-making, prediction of consequences, and handling multiple pieces of information coming into the brain, may reduce driver skills. The following signs indicate clearly that a comprehensive driver test is needed:

- Any confusion about operating the vehicle such as how to start, park or stop the vehicle.
- Any near misses such as almost hitting another car when changing lanes.
- Any side swipes even side swiping bushes on the driveway.
- Any accidents including a small fender-bender.

A person who has experienced any of the four situations just mentioned should either take a comprehensive driver test to determine ability to drive or stop driving. A comprehensive driver evaluation test may take three to four hours and usually involves tests of the: memory, cognition (thinking functions), vision, and other tests in addition to the on-the-road test.^{35,43}

Caregiver Tips

Caregivers of someone with FTLN gradually gain more responsibilities over time and must learn to conserve their energy by prioritizing tasks; they should consider limiting less enjoyable or burdensome social or other commitments. Doing what is most important and omitting unimportant chores help to conserve energy. Some fussy details in housekeeping and some burdensome friends are worth ignoring.

A sense of humor can reduce stress, refocus the attention, and keep the setting positive. It is important to observe unpleasant or stressful behaviors early and try to avoid any problems before they start. Sometimes it helps to distract quickly with a change of subject, change of pace, a louder or softer voice, slower speech (repeating simple phrases), a change of activity or offering a snack. Sometimes the caregiver just needs to leave the room for a few minutes to clear her or his head. It may be better to start another, easier task and leave the problem task for a later time. Sometimes it helps to quiet the setting, such as closing the windows, unplugging the telephone or T.V. or ushering visitors to another room, outdoors, or (their) home.

A clever way to think about providing care is to remember the word "KISSSSS". In other words, **Keep** it **Sweet** (positive), the **Same** (routine), **Simple** (priorities only, doing only what is important and preferred), **Short** (brief: activities, explanations and conversations), and **Safe** (safe: activities, setting, behaviors for everyone especially the caregiver).

A family or paid caregiver **must have respite** (take regular timeout for "down time", rest and relaxation). **Regular respite is critical!** Family caregivers, especially, should get help before actually needing it to avoid burnout or a physical collapse. If the caregiver is wondering, "Is it time to get help?", then it is time! Getting help when a caregiver is on the point of physical or emotional collapse may be too late to keep the caregiver, the family unit, the extended family and household functioning well.⁴²

In addition to receiving medical advice regarding recommendations for health care and available treatments to improve function, the family should seek advice from therapists about planning for function changes in the months or years ahead. Such planning regarding speech, occupational and other therapy evaluations should cover recommendations for the care receiver, the care provider and the setting, at home as well as other places in the community.

Speech Therapy

Speech therapy may help people with difficulties understanding or expressing language. Speech therapy may involve practicing speech, doing exercises that emphasize "errorless learning", and putting word or picture labels on the doors of rooms, closets and cabinets. Alternative strategies that supplement the lack of spoken words include picture-language books or cards (the person sees the picture and points to the word or phrase to communicate).

Amerind, (American) Indian Sign Language, which is a system of signals or gestures, may be useful to aid communication. For people who are losing language ability, Amerind may be much easier to learn than American Sign Language which includes finger-spelling and grammar rules. Examples of Amerind signs are the common gestures of 1) waving goodbye and 2) gesturing for someone to come closer. Computer programs also may help people with speech deficits.⁴⁴

People who speak slowly need more time to respond to a request. When starting an activity or after asking a question, a caregiver should allow 15 seconds or more (counting slowly and silently to 15 or 20) to provide enough time for the person to respond in a conversation.

Speech therapists also may teach people with swallowing difficulties strategies to ease swallowing and to avoid choking. For example, the person should think about the steps involved in eating and take enough time for each step. Slowing down the steps to bite, chew, swallow, clear one's throat (a cough may help), should occur before taking the next bite of food. Some foods and liquids are easier to swallow. Foods that are moister, smoother and in smaller bites are easier to eat. Liquids that are thicker are easier to swallow. In other words, water, apple juice and cranberry juice are harder to swallow while orange juice, tomato juice or soup, and apricot nectar (juice) are easier to swallow.

Occupational Therapy

An occupational therapist recommends ways or tools to simplify tasks and increase the safety of the setting. Occupational therapists offer tips that remove unnecessary steps in a task and facilitate function. An occupational therapist knows ways to simplify housekeeping and personal care tasks. Their suggestions for safety cover the home, the yard, other settings, and getting in and out of the car.

Adapting the home environment eases moving about the house, using the furniture, and doing chores. Suggestions to ease food preparation, other daily routine tasks, and personal care help the care receiver keep a sense of self-care, worthwhile purpose in daily life, self-respect, and dignity. For example, suggestions for people with frontal-temporal dementia may include laying out clothing with the first items on top such as underwear on top of the shirt, which is on top of the pants, which is on top of a sweater or jacket. Socks should be on top of the shoes which should be set on the floor by the chair where the person will sit to put on the socks and shoes.

Activity and Exercise

While a daily routine is important, activities should vary. The variety may involve personal care activities, leisure activities, quiet time alone and time involved with others, activities that use more energy and then some that are more restful, etc. Activities during the morning should involve more effort to increase the alertness and energy level of the person; activities in the evening should help the person to “wind down” as a method to begin relaxing and preparing for bedtime and good sleep.³⁵

As an example, music that is lively with a quick, bouncy beat may help energize a person; bright light (though not glaring) may help to keep a person more alert. More relaxing, slow paced music and dimmer but enough light may help a person wind down at night in preparation for sleep.

If a person is restless or agitated, soothing music, soothing light, quiet conversation, and restful activities such as reading aloud a favorite story or looking at pictures of birds or flowers may be calming.

Health professionals, social service providers and community resource people may suggest activities and daily exercises such as the following³⁵:

1. **Brain exercise** every day (any activities that use memory, language and other thinking skills): crossword puzzles, math puzzles, mazes on paper or garden mazes, number games (keeping score or mentally adding up prices for purchases); reading, watching T.V., or listening to the radio and discussing the information, looking at scenes or pictures (family photos, paintings at a store or museum, pictures in a book or on a calendar, etc.); music with words, music without words, familiar music, new music, conversations about the “old days” (reminiscing), debates about “hot” topics such as politics, etc.
2. **Walking**; short walks with occasional rest stops on a bench or chair
3. **Pet care**: brushing the dog or cat; cleaning the bird cage or bird bath
4. **Watering indoor or outdoor plants**; yard work
5. **Blowing bubbles** outdoors
6. **Washing the car**, porch furniture and yard furniture
7. **Winding yarn** or thick thread around a spool, paper tube, rolled up magazine or old tennis ball
8. **Arm, leg, shoulder, or hand massage**
9. **Simple, one-step house tasks** such as sweeping, polishing a counter top or table, vacuuming, folding laundry (towels and other easy-to-fold items)
10. **Chair exercise**:
 - a. while seated in a sturdy chair (with side support), lift arms and legs, in a series of moves. Add music or an exercise video.
 - b. march in place (feet on the floor while seated)
 - c. kick the ball
 - d. volley ball with a balloon

After the Diagnosis: Family Education and Planning Ahead

After the medical evaluation and the diagnosis, education about the condition and planning ahead are essential for long-term care management⁴². FTLD may be more challenging to deal with because of the young age of onset and the essential family and community roles of the affected person, such as employee and provider of income, support, leadership, stability, love, and innumerable services to the family and others.

Education

Education of the person with the diagnosis and the family caregivers (and significant others) will help them to understand the areas of good function versus decline. Education will help them address the changes in abilities to expect over time. Thus, they may develop strategies ahead of time or at the first sign of difficulty with a skill.^{31,35,42}

Changes in function depend upon which brain cells are affected. The type and rate of changes and the decline in ability differ from person to person. There may be “good days” and then “not-so-good-days”; later, there may be “good moments” and then “not-so-good-moments”.

Education helps the family caregivers anticipate changes in behavior or capability and modify the home schedule and setting to make self-care doable, as much as possible, during the progressive decline. Evaluations of the home setting should focus on modifying the setting rather than trying to modify the person with the diagnosis. The approach of modifying the setting may decrease caregiver stress and patient frustration.^{18,21}

Planning Ahead

As soon as possible the family should plan ahead; family matters, such as financial, legal, and health planning, should be discussed and organized^{31,35,42}. These plans should be discussed with at least one trustworthy person who does not live under the same roof as the person with the disease. A storm or other disaster that creates electrical shortages or destroys the home may lead the person with the diagnosis and the family caregiver(s) to a temporary move in another setting. If the people at this temporary setting have information about the health situation and daily care needs, it should ease the transition and the adjustment to the temporary setting.^{35,42}

It may help to set up ahead of time a “crisis carton”. The crisis carton should be a plastic (waterproof) container with a tight-fitting cover. Items to pack include copies of important health information, contact names and phone numbers, simple clothing for a few days, undergarments (disposable underwear for incontinence), family photos, paper and pens, a battery operated radio and flashlight, fresh batteries, a cell phone charger, a first aid kit, blankets, and snacks (canned liquids and dry food).

The back-up plans should consider resource people, such as family or neighbors, and resource programs and services such as local support groups and chapters of the national Alzheimer’s Association, Alzheimer Resource Centers, and local shelters that provide for people with special needs in the event of evacuations due to bad weather or other emergencies.

A *Checklist on Family Matters* is available to guide planning ahead which includes planning about money and legal matters, health preferences, organizing bill payments, and keeping forms up to date. Tasks to consider when planning ahead should include issues of safety, exercise (physical, emotional, social, and spiritual) as well as brain exercise, mobility, nutrition, physical health, dental health, personal interests such as hobbies and preferences (for example, preferences in activities and diet), and end-of-life choices.^{31,35,42}

Resources

Resources to help caregivers and care receivers with education, support groups or other services whenever appropriate. Resource programs include local chapters of the Alzheimer's Association, other groups who deal with progressive dementia such as the Alzheimer Resource Centers, adult day (health) care programs, senior centers, meal sites, religious organizations with programs or special services, assisted living facilities, nursing homes, volunteer groups and neighbors.⁴⁵

Because information on the (computer) internet is available 24 hours a day, the internet provides readily available (usually free) help. Entering in a name of a diagnosis or a medicine or a challenging behavior will result in a wealth of explanations and suggestions. Some excellent internet web sites and toll free phone numbers are listed in the following Table 1.

Table 1. Information on FTLD: Internet sites and Toll Free Phone Numbers
<p>▶ Association for Frontotemporal Dementias: www.ftd-picks.org</p> <p>▶ National Aphasia Association: www.aphasia.org; (800)922-4622</p>

When considering helpers, the list should include not only health and social service providers but also care managers who help address the wide range of daily and weekly needs of the family and the household. The family needs to recognize that a person with a frontotemporal lobar degeneration may be an unsafe driver; poor judgment, faulty decision-making, and slow, stiff movement increase the risks for accidents when operating a motor vehicle. Planning ahead for alternative transportation should occur before it is clear (because of an accident or near miss) that the family unit no longer has a safe or available driver.⁴³⁻⁴⁸ Important resources on driving safety are listed in the following Table 2.

Table 2. Some Internet Resources on Driving Issues and Safety⁴⁶

- ▶ **Alternative transportation:** <http://www.eldercare.gov>
- ▶ **American Occupational Therapy Association:** <http://www.aota.org>
- ▶ **Association of Driver Rehabilitation Specialists:** <http://www.aded.net>;
- ▶ **CarFit:** www.eldersafety.org/latest/roadmap-to-driving-wellness-and-car-fit-two-exciting-new-programs.html
- ▶ **Driver refresher courses:** <http://www.aarp.org/drive>; www.aaapublicaffairs.com
- ▶ **Driving & Dementia Toolkit (published in Journal of the American Geriatric Society):** <http://rgapottawa.com/dementia/english/default.asp>
- ▶ **DriveWell:** <http://www.asaging.org/drivewell>
- ▶ **GrandDriver information & references:** <http://www.granddriver.info/>
- ▶ **Independence Drive:** <http://driving.phhp.ufl.edu/index.php> (NODRTC website); <http://www.phhp.ufl.edu/ot/> (OT website); <http://www.IndependenceDrive.phhp.ufl.edu>
- ▶ **Older driver facts:** <http://www.seniordrivers.org>; www.nhtsa.dot.gov; www.aoa.gov

Other resources listed in the following Table 3 provide general information about progressive dementias and caregiver management that may be of help to people dealing with frontotemporal lobar degeneration.

Table 3. General Resources

- ▶ Alzheimer's Association (USA): www.alz.org or www.alzheimers.com; (800)272-3900
- ▶ Alzheimer Society of Canada: www.alzheimer.ca
- ▶ Alzheimer's Society (United Kingdom): www.alzheimers.org.uk
- ▶ AlzOnline (at University of Florida): www.AlzOnline.net
- ▶ American Academy of Neurology: www.aan.com; www.thebrainmatters.org
- ▶ Family Caregiver Alliance: www.caregiver.org; (800)445-8106
- ▶ Helpguide, Aging Issues: www.helpguide.org/elder/lewy_body_disease.htm
- ▶ Los Angeles Caregiver Resource Center: <http://geroweb.usc.edu/lacrc>; (800)540-4442
- ▶ National Institute of Neurological Disorders and Stroke: www.ninds.nih.gov/disorders

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