What is Frontotemporal Dementia?

Frontotemporal dementia (FTD) is a progressive, degenerative brain disease that gradually destroys the ability to behave appropriately, empathize with others, learn, reason, make judgments, communicate and carry out daily activities. In people under age 60, FTD is the most common cause of dementia and affects as many people as Alzheimer's disease in the 45-64 age group. Men are affected more commonly than women. There are several forms of the disease that lead to slightly different behavioral, language and/or motor symptoms. Due to the symptoms, FTD can be mistaken for Alzheimer's disease, Parkinson's disease or a primarily psychiatric disorder like depression, manic-depression, obsessive-compulsive disease or schizophrenia. There is no treatment or cure yet that can reverse the damage, but medications and lifestyle changes can help relieve the symptoms. FTD is not contagious.

Forms of Frontotemporal Dementia

Based on the distinct patterns of signs and symptoms, three different clinical syndromes have been grouped together under the category of "frontotemporal dementia" (FTD):

1. Behavioral-variant frontotemporal dementia (bvFTD)
2. Semantic dementia (SD) and

Previously, researchers sometimes used "FTD" to refer only to bvFTD, which has also been called "frontal variant FTD" (fvFTD) or Pick's Disease. The language variants (SD and PNFA) are sometimes grouped together under the term "primary progressive aphasia" (PPA). PPA has since been split into three subgroups: progressive non-fluent aphasia, semantic dementia and logopenic progressive aphasia (LPA). At autopsy, patients with LPA are often found to have Alzheimer's disease, not FTLD. SD was previously referred to at times as "temporal variant FTD" (tvFTD).

A small number of people affected by FTD also develop motor neuron disease (FTD/MND), (sometimes called FTD with amyotrophic lateral sclerosis or FTD/ALS).

Corticobasal degeneration (CBD), also called corticobasal syndrome or corticobasal ganglionic degeneration, and progressive supranuclear palsy (PSP) are two related diseases that are not classified as FTD but often share some symptoms with FTD.

Behavioral variant FTD

Behavioral variant frontotemporal dementia (bvFTD) has also been referred to as "frontal variant FTD" (fvFTD) or "Pick's disease." Approximately 60% of people with any form of FTD have bvFTD. By definition, this form of FTD affects social skills, emotions, personal conduct, and self-awareness. Deficits in these functions most often reflect damage to specific regions within the frontal and temporal lobes. With damage to these areas, people may show mood and behavior changes including stubbornness, emotional coldness or distance, apathy and selfishness. Unlike Alzheimer's disease, which affects a different area of the brain, many
people with bvFTD don't show any confusion or forgetfulness about where they are or what day it is, at least at first.

Semantic dementia

Semantic dementia, which has also been called "temporal variant FTD," accounts for 20% of FTD cases. Language difficulty, the predominant complaint of people with SD, is due to the disease damaging the left temporal lobe, an area critical for assigning meaning to words. The language deficit is not in producing speech but is a loss of the meaning, or semantics, of words. At first, you might notice someone substituting a word like "thingy" for more unusual words, but eventually a person with SD will lose the meaning of more common words as well. For example, early in the illness a patient might lose the word for a falcon, later-on forget the word for a chicken, then call all winged creatures "bird" and eventually call all animals "things." Not only do they lose the ability to recall the word, but the concept of these words is also lost. "What is a bird?" might be a typical response for a patient with advanced SD. Reading and spelling usually decline as well, but the person may still be able to do arithmetic and use numbers, shapes or colors well. Names of people, even good friends, can become quite difficult for people with SD. Like the behavioral variant, memory, an understanding of where they are, and sense of day and time tend to function as before. Muscle control for daily life and activities tends to remain good until late in the disease. Some of these skills may seem worse than they actually are because of the language difficulty people with SD have when they try to express themselves.

When SD starts in the right temporal lobe, people in the early stages have more trouble remembering the faces of friends and familiar people. Additionally, these people show profound deficits in understanding the emotions of others. The loss of empathy is an early, and often initial, symptom of patients with this right-sided form of SD. Eventually people with right-sided onset progress to the left side and then develop the classical language features of SD. Similarly, left-sided cases progress to involve the right temporal lobe and then the person experiences difficulty recognizing faces, foods, animals and emotion. SD patients eventually develop classical bvFTD behaviors including disinhibition, apathy, loss of empathy and diminished insight. The time from diagnosis to the end is longer than for those with bvFTD, typically taking about six years.

Progressive nonfluent aphasia

PNFA accounts for only about 20% of all people with FTD. Unlike semantic dementia where the person maintains the ability to speak but loses the meaning of the word, people with PNFA have difficulty producing language fluently even though they still know the meaning of the words they are trying to say. The person may talk slowly, having trouble saying the words, and have great trouble with the telephone, talking within groups of people or understanding complex sentences. In recent years it has become apparent that many patients with PNFA go on to develop severe Parkinsonian symptoms that overlap with progressive supranuclear palsy (PSP) and corticobasal degeneration (CBD) such as an inability to move the eyes side-to-side, muscle rigidity in the arms and legs, falls, and weakness in the muscles around the throat.

FTD with motor neuron disease

Approximately 15% of patients with FTD also develop motor neuron disease (FTD-MND). Most often, this combination occurs in patients with bvFTD, and only rarely does MND arise in patients with SD or PNFA. MND affects motor nerve cells in the spinal cord, the brain stem (which sits on top of the spinal cord), and the cerebral cortex. Because the brainstem was once referred to as the “bulb”, you may hear some MND symptoms described as "bulbar symptoms". The most common type of MND is amyotrophic lateral sclerosis (ALS), also called Lou Gehrig's disease, which can occur as a purely motor disorder. More often, however, patients with ALS also have behavioral or cognitive problems similar to those seen in FTD. MND symptoms include slurring of speech, difficulty swallowing, choking, limb weakness or muscle wasting. In patients with FTD-MND, there
is often (but not always) a family history of the disease, and scientists are getting closer to identifying gene
mutations that cause the illness.

How Do You Know if it's FTD?

In trying to determine what is happening to a person, a doctor must first review the important signs and
symptoms with the patient and caregivers. Determination of the neuropsychological, neurological, psychiatric,
general medical and functional status of the patient is also important. People with FTD are often misdiagnosed
with Alzheimer's disease, psychiatric problems, vascular dementia or Parkinson's disease. MRI can help rule out
other diseases and support a diagnosis of FTD.

Could it be Alzheimer's?

Alzheimer's disease (AD) is the most common dementia in older people. Therefore, it should be one of the first
diseases your doctor considers. Alzheimer's disease usually begins with memory loss while FTD is usually a
behavior or language disorder.

People with either disease will show cognitive difficulties and multitask poorly. And at the end stages, AD and
FTD look very similar. Doctors use the early symptoms and the brain image, usually done on a MRI (magnetic
resonance imaging) scanner, to reach the most appropriate diagnosis.

- The probability of AD is strongly affected by the age of the person showing the symptoms. The odds of
  having Alzheimer's disease increase markedly the older you get while the odds for FTD may decrease
  with age.
- FTD often begins with distinct behavioral changes (socially inappropriate, apathetic, impulsive, etc.)
  while people with Alzheimer's in the early stages tend to remain socially graceful despite their memory
  problems (they may even become skilled at covering up their difficulties). In advanced AD, people
generally have trouble managing their finances, show poor judgment and irritability, and may become
equally difficult to manage as FTD.
- Apathy in AD patients is milder, whereas apathy in FTD patients is more pervasive and more often
  reflects a lack of concern for others or lack of initiative.
- AD patients have an early and profound difficulty learning and retaining new information. As the
disease progresses, memory for new and old information is lost. These memory problems may lead to
language problems as well, but the root is a problem remembering. In contrast, most mildly impaired
FTD patients generally know the day or time and their location, and they are able to keep track of recent
events. They may not test well, but that may be due to lack of concern or effort in the testing situation.

Could it be a psychiatric problem?

When behavioral symptoms predominate, FTD patients who become ill in mid-life may be confused with
patients who have late life depression, or when the onset is in younger persons, the FTD may be confused with
schizophrenia or bipolar disorder. Repetitive compulsive behaviors are very common in bvFTD, and some
patients may initially be given the diagnosis of obsessive-compulsive disorder. Since the history and exam may
look very similar for a psychiatric patient and an FTD patient, neuropsychological testing and a brain image will
help clarify the picture.

Confirming an FTD diagnosis
Once other likely diseases have been ruled out, an FTD diagnosis is made by looking at the data from a neurological exam and personal history (which may come from the patient, family or other caregiver); neuropsychological tests that help quantify memory, language and other cognitive skills; and a brain image - usually a MRI (magnetic resonance imaging) scan but perhaps a functional scan like PET (positron emission tomography) which can show increased or lowered brain activity in the frontal and anterior temporal areas.

An accurate diagnosis made after ruling out other possible explanations is essential for successful treatment or management of any disease. As new medical treatments become available, early intervention will be more and more important. Regardless of medical treatments, the sooner patients and caregivers have an accurate diagnosis, the sooner they can plan for upcoming care. As FTD tends to affect younger people than Alzheimer's or other dementias and lasts for eight years on average, creating a care plan is particularly important for everyone touched by this disease.

**Signs and Symptoms**

People with FTD typically first come to the doctor's office because of:

1. Gradual and steady changes in behavior,
2. Gradual and steady language dysfunction or
3. Gradual and steady weakness or slowing of movement

**Behavioral symptoms**

Apathy is often the first symptom reported by caregivers and may be mistaken for depression. People experiencing these changes may become self-centered, emotionally distant, withdrawn, unaware of the emotions of others, avoid social contact or neglect previous hobbies and interests. They may develop a lack of concern for their personal appearance and become increasingly unkempt early in the course of disease.

Impulsive behavior is another common complaint from caregivers who may find the changes in social and personal conduct embarrassing or frustrating. These behaviors are often associated with a lack of inhibition, resulting in impulsive or inappropriate behavior, such as overeating, outbursts of frustration, touching strangers, urinating in public or diminished social tact. Overeating is common and "food fads" can occur where the person with bvFTD will only eat certain foods. Caregivers often notice an overactive "sweet tooth." Restlessness, irritability, aggressiveness, violent outbursts or excessive sentimentality is not unusual either.

There is usually difficulty in reasoning, judgment, organization and planning, and consequently, these patients can be quite gullible and fall prey to scams on the computer or in person. As the disease progresses, this lack of judgment may lead to criminal behavior (such as shoplifting, indecent exposure, running stop lights, poor financial judgment or impulsive buying). At the extreme, the impulsivity can be self-destructive, as when patients try to get out of a moving car. In some people, inappropriate sexual behavior occurs.

There may also be repetitive or compulsive behaviors that may include hoarding, doing the same thing over and over (for instance, reading the same book several times or walking to the same location again and again), pacing, or repeating particular "catch phrases" over and over in their speech.

The person with bvFTD may experience false thoughts (delusions) that are jealous, religious or bizarre in nature. Or they can develop euphoria - excessive or inappropriate elation or exaggerated self-esteem.

Even though they might complain of memory disturbance, patients with the behavioral variant of frontotemporal dementia can usually keep track of day-to-day events and understand what is going on around
them. Also, for people with bvFTD, their language skills and memory usually remain intact until late in the disease.

These behaviors have a physical cause and are not something that the person can usually control or contain. Indeed, often the person has little or no awareness of the problem behaviors.

Language symptoms

The majority of patients with the language variants have problems expressing themselves with language while their memory stays relatively intact. Difficulties reading and writing then develop. To understand more about language, see our Speech and Language section.

At the UCSF Memory and Aging Center, we have found a small group of FTD patients who develop new creative skills in music and art as their language skills decline. For more information about this topic, please see this article on Personality and Creativity.

Semantic dementia (SD)

The most common complaint of people with semantic dementia (SD) is increasing trouble naming people, objects, facts and words. As the disease progresses, they lose not only the ability to name something, but also the meaning of what it is they are trying to name – like how to use it or to what context it belongs. People with SD usually know they are having trouble finding their words and understanding what is being said to them. Their speech tends to keep the usual speed and rhythm, but they may substitute similar but incorrect words or replace a word with "thing" or "stuff." Patients continue to speak the same amount, even as the disease progresses. Some people may develop an inability to recognize familiar faces. Later in the disease course, similar behavioral changes to those seen in bvFTD may appear.

Progressive non-fluent aphasia (PNFA)

People with PNFA tend to come to the doctor's office with complaints about changes in their fluency or rhythm of speech, pronunciation or word finding difficulty. These patients tend not to show the behavioral characteristics of FTD until quite late in the disease, and they are keenly aware of their difficulties. Depression and social withdrawal are common features of PNFA. As the disease progresses, less and less language is used, until the patient may be virtually mute.

Motor symptoms

People with FTD often describe a general weakening of their muscles or slowing of their movements. They might feel uncoordinated or like they are walking through water - harder to move and slower going. They may also experience muscle spasms. In a neurological exam, the doctor may also find some slowing of particular eye movements, changes in the typical reflexes and muscle stiffness or slowness.

Disease Progression

While each type of FTD produces different symptoms, all the types cause a steady decline in the ability to think and function, eventually leaving the person completely dependent on caregivers to get through the day. The disease usually first appears when someone is in their mid-40s to early-60s and lasts an average of eight years.

Behavioral variant FTD
The pace of the symptoms and length of disease can vary dramatically from person to person. In general, each type of FTD follows a pattern where the symptoms seen in the mild stage become more pronounced and disabling over a course of 8-10 years.

**Mild bvFTD**

In the first several years, a person with bvFTD (sometimes called Pick's disease or just FTD) tends to exhibit marked behavioral changes such as disinhibition, apathy, loss of sympathy or empathy for others, or overeating. Problems with planning organization and sometimes memory are evident, but the individual is still capable of managing household tasks and self-care with minimal help. However, impairment in judgment can lead to financial indiscretions with potentially catastrophic consequences. Social withdrawal, apathy and less interest in family, friends and hobbies may be evident. At times, they may behave inappropriately with strangers, lose their social manners, act impulsively and even break laws. But at this stage, the behaviors can often be managed with lifestyle and environmental changes (read our practical tips for ideas). A MRI image at this point will show mild atrophy in particular areas of the frontal lobes.

**Moderate bvFTD**

Over the course of a few years, the symptoms seen in the mild stage will become more pronounced and disabling. You might also notice compulsive behaviors like repetitive urination, hoarding or collecting objects, compulsive cleaning or silly repetitive movements (like stomping on ants). Binge eating may create weight problems and other health issues. The cognitive problems associated with dementia become more pronounced, with mental rigidity, forgetfulness and severe deficits in planning and attention. The MRI image at this point will show that the shrinking of the brain tissue has expanded to larger areas of the frontal lobes, as well as the tips of the temporal lobes and basal ganglia, deeper brain structures involved in motor coordination, cognition, emotions and learning.

**Severe bvFTD**

By this point the patient is experiencing profound behavioral symptoms (apathy, loss of empathy, disinhibition) in association with language difficulty and memory loss. Although it can vary widely, the time from the first symptom to the end is typically about eight years, whereas the time from diagnosis is, on average, about five years.

**Semantic dementia**

The pace of the symptoms and length of disease can vary dramatically from person to person. In general, each type of FTD follows a pattern where the symptoms seen in the mild stage become more pronounced and disabling over a course of 8-10 years.

**Mild SD**

People with early semantic dementia that is predominantly on the left side of the brain, usually complain of a hard time coming up with the word or name for something. Words that the person uses a lot may remain available, but more unusual words may be replaced by "thingy" or "you know." The tone, rhythm and fluctuations of pitch (prosody) generally sound normal. Memory for day-to-day events is usually spared.
The early signs of SD in people with asymmetric right-sided damage include a decline in empathy or awareness of other people's emotions.

**Moderate SD**

After two to three years, the people with left sided damage and those with right sided damage tend to look more similar, as the disease typically progresses to involve both sides. With moderate SD, most people show at least some of the behavioral problems that are similar to the behavioral variant of FTD.

People with moderate semantic dementia will have immense trouble understanding you. They may also have increasing difficulty recognizing the names and faces of people – even friends and family. Reading and writing, mostly likely, will have declined noticeably. The person may still be able to use numbers, colors and shapes – the brain functions responsible for these skills are organized in a different area of the brain from words.

**Severe SD**

After four to five years of SD, the disease is usually quite advanced, which means the person's language skills have significantly eroded, making communication very difficult while the behavioral problems have significantly increased. Typical behaviors seen in late stage SD include disinhibition, apathy, compulsions, impaired face recognition, altered food preference and weight gain. People with left-sided damage tend to show more interest in visual or non-verbal things while people with right-sided damage tend to prefer games with words and symbols. The time from diagnosis to the end typically takes about six years, although this can vary significantly from person to person.

**Progressive nonfluent aphasia**

The pace of the symptoms and length of disease can vary dramatically from person to person. In general, each type of FTD follows a pattern where the symptoms seen in the mild stage become more pronounced and disabling over a course of 8-10 years.

**Mild PNFA**

Progressive nonfluent aphasia leads to increasing trouble speaking and producing language, although the person with it usually understands language and knows what they want to say. Early symptoms include slowed speech and trouble getting the works out correctly. For example, if a person with PNFA has to repeat a word that is difficult to say several times, it will most likely sound a little different each time.

**Moderate PNFA**

As the disease progresses into years three and four, the person will have more and more trouble producing speech. They may use short sentences without a lot of extra words like articles and adjectives. Reading and writing skills may are usually still good, so you might want to consider using a board or number of pictures to help the person express their meaning. Skills with numbers, colors and shapes generally remain intact, as do skills involving face and emotion recognition.

**Severe PNFA**

After five or more years of PNFA, the person with PNFA is essentially mute and may show behavioral problems similar to those of behavioral variant FTD. Some people with PNFA develop Parkinson's-like motor problems like muscle rigidity and stiffness. The time from diagnosis to the end typically takes about six years, although this can vary significantly from person to person.
End stage FTD

After years of FTD, patients may have trouble coordinating their muscles and require a wheelchair. Usually 24-hour care is required, whether at home or in an institution. The physical decline and changes that occur throughout the disease course become more and more obvious at this stage. Eventually, the person with FTD may have great difficulty swallowing, chewing, moving and controlling their bladder and/or bowels. Death from FTD is usually caused by the consequences of these physical changes, most commonly infections in the lungs, skin or urinary tract. Although it can vary widely, the time from the first symptom to the end is typically about eight years, whereas the time from diagnosis is, on average, about five years.

Treatments

Unfortunately, there is no way to reverse the damage caused by FTD yet, but many medications and lifestyle changes can help relieve the symptoms. Furthermore, researchers are actively searching for new treatments and running clinical trials to test promising new medications.

Diagnosis

The first step to treatment of any illness is getting the correct diagnosis. While there are currently no treatments available to stop or reverse frontotemporal dementia, getting the right diagnosis prevents you from receiving incorrect or potentially harmful medications and also provides an opportunity to join a clinical trial where you may be eligible for new treatments.

As part of the initial evaluation, your doctor should carefully review all your medications - prescription, non-prescription (over-the-counter), alternative, nutritional supplements and recreational drugs such as alcohol, caffeine, tobacco, etc. Sometimes a bothersome symptom can be explained by a drug side effect or drug-drug or drug-disease interaction.

Lifestyle and environmental changes

Once you have a diagnosis, as a patient or caregiver you should ask your doctor for resources where you can learn more about the disease and find others who are going through a similar experience. Frontotemporal dementia, particularly the behavioral type, can create safety issues in the home and around driving. Patients might leave pots on the stove, wander outside while inappropriately dressed, get lost, have aggressive outbursts or exhibit other potentially risky behaviors. You may need to remove dangerous items from the home and take away driving privileges.

Often times, adjusting expectations and making changes to the environment can help improve the patient's behaviors. Learning to roll with disruptive but non-dangerous behavior takes patience, but sometimes saving your energy for more serious matters is the best strategy. Review our Practical Tips section for ideas to help manage symptoms and daily routines. Support groups are another great way to learn non-medical ways to manage the disease - other caregivers often have great ideas.

End-of-life planning should begin as soon as there is a diagnosis so that everyone can participate. The documents can always be updated or changed at a later date, but it helps everyone to begin the discussions early and create at least simplified legal documents.
An exercise program that combines both strength training and aerobics is important for maintaining cognitive health. Cardiovascular exercise 2-4 days/week for approximately 30 minutes each time is beneficial. A regular walk is one form of cardiovascular exercise and does not require membership to a gym. A common saying at the Memory Aging Center is, “What is good for the heart is good for the brain.” Individuals should consult with their physician before beginning any exercise program. Regular exercise can help you and your loved one maintain your ability to function for a longer time. Don't only think about the patient getting exercise; consider a formal regimen for yourself. Getting enough sleep also helps maintain optimal cognitive performance.

**Speech therapy**

People with either of the language variants may benefit from speech therapy to help them adjust to their language difficulties and learn alternate ways to communicate. Maintaining adequate communication can decrease frustration, a common problem in FTD. Unlike many people who develop aphasia from head injury or stroke, people with FTD will not improve with time, but a therapist may be helpful in maximizing existing abilities and exploring other ways to communicate. Non-verbal techniques for communicating, such as gesturing, pointing to pictures, etc., may help people with SD or PNFA express themselves. Aphasia identification cards explaining that the person has a language problem can aid in communicating the person's condition to others. Many speech pathologists and occupational therapists have their own practices, while others are available through local hospitals and medical centers. Ask your doctor for a referral.

**Medications for behavioral symptoms**

Medications for behavioral symptoms should be started at a low dose and then increased slowly based on the patient’s response and the presence of side effects. It is only fair to note that very few clinical studies exist examining the benefits of the following medications, and the following information is base primarily on clinical experience.

**Antidepressants**

While originally created to treat depression, certain antidepressants are effective in treating anxiety disorders, preventing panic attacks and obsessive compulsive behavior. One group of antidepressants called selective serotonin reuptake inhibitors (SSRIs) may be useful in reducing the aggressive impulses, poor impulse control and carbohydrate craving associated with early Pick’s/FTD. People with FTD usually do not experience many side effects with SSRIs, and they are generally considered the best available medications for controlling problematic behaviors.

Examples of SSRIs include:
- fluoxetine (Prozac®)
- sertraline (Zoloft®)
- paroxetine (Paxil®)
- fluvoxamine (Luvox®)
- citalopram (Celexa®)
- escitalopram (Lexapro®)

Other antidepressants (not SSRIs) that may be useful:
- trazodone (Desyrel®)
- venlafaxine (Effexor®)
- duloxetine (Cymbalta®)
- bupropion (Wellbutrin®)
mirtazepine (Remeron®)

Antipsychotics

Antipsychotic medications block the effects of dopamine, a chemical messenger that can increase hallucinations, delusions (false beliefs) and can alter rational thought. Low doses of these medications can help manage aggressive, irrational and compulsive behaviors that may develop with frontotemporal dementia. The potential benefit of antipsychotics must be weighed against potential risks including weight gain, slowing of movement and thinking, accelerating heart disease and, in rare instances, death. Typical antipsychotics are associated with muscle problems and should be avoided, since patients with FTD are likely to show muscle stiffness and trembling.

Examples of atypical antipsychotic medications include:
- olanzepine (Zyprexa®)
- quetiapine (Seroquel® or Ketipinor®)
- risperidone (Risperdal®)
- ziprasidone (Geodon®)
- aripiprazole (Abilify®)
- paliperidone (INVEGA®)

Other classes of drugs that might be useful, but have not been studied in Pick's/FTD are anti-anxiety drugs and anti-seizure medications used as “mood stabilizers.”
- valproic acid and divalproex sodium (Depacon™, Depakene®, Depakote®, Depakote® ER)
- carbamazepine (Tegretol®)
- gabapentin (Neurontin®)

Medications for memory

Memantine (Namenda®), a NMDA-receptor antagonist, is approved for treating Alzheimer's. Preliminary evidence suggests it may provide some benefit for people with Pick's/FTD as well. The Memory and Aging Center will soon complete a research trial on memantine.

Medications to avoid

Cholinesterase inhibitors, such as donepezil (Aricept®), rivastigmine (Exelon®) and galantamine (Razadyne®), are prescribed for people with Alzheimer's disease. They seem to work by helping people to improve attention and working memory by increasing the amount of acetylcholine, a chemical in the brain. Unfortunately most people with frontotemporal dementia do not benefit from these medications and may even become more agitated when they are tried. Generally, cholinesterase inhibitors are not recommended for patients diagnosed with FTD.

Benzodiazepines, used to treat anxiety, insomnia, agitation, seizures and muscle spasms, have been linked to increased behavioral problems and impairment in both memory and psychomotor skills.

Common benzodiazepines:
- estazolam (ProSom®)
- diazepam (Valium®)
- flurazepam (Dalmane®)
- midazolam (Dormicum®, Versed®)
- temazepam (Restoril®, Normison®)
- triazolam (Halcion®)
• alprazolam (Xanax®)
• chlordiazepoxide (Librium®)
• clonazepam (Rivotril®, Klonopin®)
• clorazepate (Tranxene®)
• lorazepam (Ativan®, Temesta®, Tavor®)
• oxazepam (Serax®, Serepax®)
• prazepam (Centrax®, Lysanxia®)

Typical antipsychotics such as haloperidol (Haldol®), fluphenazine (Prolixin®) and chlorpromazine (Thorazine®) are associated with muscle problems and should be avoided, since patients with FTD are likely to show muscle stiffness and trembling from the disease.

If you are concerned about any of the medications you are taking, ask your doctor or pharmacist to review them for you. It is always a good idea to take all your medications with you to each doctor's appointment so the types and dosages can be reviewed.

RxList and Healthline provide helpful online tools for checking potential medication side effects and interactions.

To Discuss With Your Doctor

A good relationship and clear communication with your doctor will result in the best care for the person with FTD. Learning all you can about FTD will help you be prepared to discuss symptoms, tests and opportunities to participate in research. Feel free to download our Frontotemporal Dementia Primer that you can give to your primary care physician in case they have not treated someone with FTD.

Talking With Your Doctor

Being prepared when you come in for an office visit can help you get your concerns addressed and produce a more accurate diagnosis earlier.

Communication Tips

If you or someone you know is experiencing any of the signs and symptoms of FTD, you may want to talk to a doctor about your concerns and observations.

A good relationship and clear communication with the doctor will result in the best care for your loved one. Here are some tips to help make talking to your doctor more effective:

- **Be prepared.** Several days before your appointment, use these worksheets to help you track medical history, health changes, your concerns and current medications. When your appointment time comes, arrive early to fill out forms, bring two copies of your completed worksheets so you can go over it together and bring a pen and paper to jot down notes.
- **Speak up.** Doctors tend to prioritize diagnostic information and core concerns early in the office visit. Mention your most important concerns early in the visit to be sure you have time to cover them. It might be helpful to use the list in the appointment worksheets.
- **Listen.** Sometimes it feels like a doctor's appointment is over in a blink of an eye and all you walk out with is the blurred memory of a meeting and a prescription. Take notes or bring a tape recorder and ask the doctor if she wouldn't mind your recording the visit to help you better remember the information you discuss.
- **Ask questions.** Don't hesitate to ask for clarification if your doctor uses unfamiliar words or gives confusing instructions. Any recommendations you get need to fit your personal, cultural and financial situation so that you can put them into practice. Don't leave the office without understanding everything the doctor tells you.

**Starter Questions You Might Ask Your Doctor**

Here are some questions you might want to ask your doctor or nurse. Feel free to add your own questions.

1. Do you know what is wrong? How do you know?
2. What if it’s FTD? Where can I go for information, advice and resources?
3. Should I see a specialist?
4. Do I need a follow-up visit?
5. What tests do I need and why?
6. What do the tests involve?
7. Are there changes I should watch for?
8. When should I call you?

Realize that unfortunately FTD is often mistaken for other diseases, and many doctors have little experience diagnosing or managing FTD. It may be helpful to let doctors know about this website or download this Primer on Frontotemporal Dementia before your next visit.

**Tests**

Certain tests can help your doctor rule out or support a diagnosis of FTD.

**Neurological exam and history**

Because memory loss, behavioral disorders and other related problems are often complex, a comprehensive evaluation is necessary. At the UCSF Memory and Aging Center, patients undergo an extensive neurological, neuropsychological and nursing assessment that usually requires several hours. Information from the caregiver is sought in every case. The evaluation may require two to three visits to determine the cause of the symptoms and recommend treatment.

After the evaluation, the medical team involved with each patient meets to discuss the diagnosis and potential treatments. After this meeting, the team discusses its findings with the patient and the family. In some cases, a diagnosis will be deferred until more information from blood tests or brain imaging is collected.

**Neuroimaging**

**Structural scans**

One of the most useful tests in the evaluation of FTD is magnetic resonance imaging (MRI). MRI uses magnetic fields and radio waves, without any X-rays, to produce images of the inside of your body. It is non-invasive and considered very safe, but some people with metal implants and cardiac pacemakers are unable to have MRI.
Talk to your doctor or the imaging technician if you have any concerns about entering the magnet. Some people find lying in the scanner produces anxiety or claustrophobia because of the tube-like shape or the loud sounds during the scan. Sedation may be available to you if needed, but relaxation techniques like deep breathing, visualization and meditation can also help. Some MRI scanners allow you to listen to music or watch a movie. To get the best pictures, you need to be as still as possible while in the scanner.

FTD leads to loss of brain tissue that is visible on MRI scans of patients while patients are still alive. Different areas of the brain are affected (early on) by different types of FTD:

1. **FTD (frontal lobes):** responsible for personality, judgment and planning/organization
2. **SD (anterior temporal lobes):** store general information about the world, very important for language and face recognition, critical for understanding the emotions of others
3. **PNFA (left frontal lobe):** produces spoken language

A computed tomography (CT) scan is similar to the MRI but does not image brain structure with the fine precision of MR. A CT scan is an X-ray technique that produces cross-sectional images of the inside of your body or head. Typically scans last only a few minutes, during which time you should lie still. You may hear some whirring and clicking noises during this test, which is normal. In order to make the CT image, you will be briefly exposed to X-ray radiation, so be sure to discuss any concerns you have with your doctor.

**Functional scans**

Functional scans, by single-photon emission computed tomography (SPECT), functional MRI (fMRI) or positron emission tomography (PET), typically demonstrate decreased activity in the frontal and temporal lobes. Amyloid imaging with PET can tell whether the patient is suffering from Alzheimer's disease, versus frontotemporal dementia. This is still experimental but can be obtained in some centers.

A SPECT scan shows how blood flows through arteries in the brain. A radioactive material (tracer) is injected into a vein in the arm, and then the scanner detects the movement of the tracer through the brain and computes the brain activity. Brain areas affected by FTD show diminished activity. As with any neuroimaging procedure, you will need to lie as still as possible so that the machine can obtain accurate pictures. After the scan, be sure to drink plenty of fluids. Most of the radioactive tracer leaves your body through your urine within a few hours after your SPECT scan. Talk to your doctor if you're concerned about your exposure to radiation during a SPECT scan.

Functional MRI is a special type of scan done in the MRI scanner. It shows changes in blood flow in the brain, which represent active areas of the brain using more or less blood to perform certain tasks. The experience and equipment is similar to that of a structural MRI scan.

PET scans show the activity of tissues by measuring the energy usage (metabolic activity) of your brain. Like a SPECT scan, PET combines a brain camera and a radioactive material (tracer). The tracer is what allows doctors to see how your body tissues absorb and use different chemicals in real time. 30-45 minutes prior to the scan, a tracer is injected into your bloodstream. Once the tracer has had time to reach your brain, you'll lie on a table that moves slowly through the scanner. By detecting metabolic changes in the brain, your doctor can see which areas are healthy versus dysfunctional. Be sure to remain as still as possible so that the machine can get accurate pictures. Depending on the information your doctor needs, you may be asked to perform certain tasks like read or speak to activate specific areas of your brain. Once the scan is complete, be sure to drink plenty of fluids to flush out any tracer left in your body.

**Neuropsychology**
Neuropsychological testing is useful to obtain a clinical assessment of the disease. These tests evaluate conduct, language, visuospatial abilities, memory, abstraction, planning and mental control, motor skills and intelligence. Tests of the FTD patient may show visual and memory abilities intact. However, abstract thinking, word generation, motivation and ability to follow rules may be disrupted.

**EEG**

An electroencephalogram (EEG) shows patterns of electrical activity produced by your brain as recorded by electrodes placed on your scalp. It is non-invasive and minimally uncomfortable (the electrodes may scratch or itch you and are held in place with a sticky paste). The electrodes do not generate any electricity; they only record electrical activity produced by your brain. You will need to be still with your eyes closed during the 20-40 minute recording in order to get a quality EEG.

In people with FTD, the electroencephalogram (EEG) is usually normal or shows mild frontal slowing. Thus, a normal EEG does not mean that the behavioral manifestations are primarily the result of a psychiatric illness.

**Genetic Counseling**

If your family has a history of dementia or motor neuron disease, you may want to meet a genetic counselor to help assess your risk of a genetic disorder and weigh the medical, social and ethical decisions surrounding genetic testing.

**What is genetic counseling?**

Genetic counseling is the process of helping people understand and adapt to the medical, psychological and familial implications of genetic contributions to disease. The process integrates the following elements:

- Interpreting family history and medical information to assess risk of disease occurrence or recurrence
- Education about inheritance, testing, management, prevention, resources and research
- Counseling to promote informed decision-making and adaptation to the risk or condition

Trained genetics professionals include doctors who specialize as medical geneticists, genetic counselors (health professionals with specialized graduate degrees) and genetic nurses.

If you are concerned about hereditary disease, you may benefit from a genetic consultation, an important part of the decision-making process for genetic testing.

**What to expect in your consultation**

A genetic consultation provides information, addresses a patient’s specific questions and concerns, and offers support.

During a consultation, a genetics professional will:

- Assess the risk of a genetic disorder by researching your family's history and evaluating medical records.
- Help you weigh the medical, social and ethical decisions surrounding genetic testing to help you make informed, independent decisions about your health care.
- Interpret the results of genetic tests and medical data.
• Offer supportive counseling and assist you in coping with difficult situations and feelings.
• Respect your individual beliefs, traditions and feelings.
• Serve as patient advocates.
• Discuss reproductive options.
• Explore strategies for communicating information to others, especially family members who may be at risk.
• Provide written materials and referrals to support groups, other families with the same or similar condition, and local and national service agencies.

A genetics professional will not:

• Tell you which decision to make.
• Advise you not to have children.
• Recommend that a woman continue or end a pregnancy.
• Tell you whether to undergo testing for a genetic disorder.

What is genetic testing?

Genetic testing identifies changes in DNA, RNA, genes, chromosomes or proteins – the blueprint which instructs how we grow and develop. The result of a genetic test can often confirm or exclude a suspected genetic condition or help determine a person's chance of developing or transmitting a genetic disorder. Unlike routine blood tests, the results from a genetic test will not change over time and only need to be performed once in a person's life.

Approximately 20-50% of all individuals with frontotemporal dementia (FTD) have a known family history. This means that 50-80% of those affected have no known family history of the disorder. Sometimes, this is due to a misdiagnosis in the family with another condition, such as Parkinson disease, Alzheimer's or psychiatric illness. For this reason, individuals in the family may be at-risk for a "familial" disorder without prior knowledge. Genetic counseling and testing can help clarify this risk.

Genetic testing can be performed to confirm a suspected diagnosis of FTD, assess risk for extended family members or identify an individual with increased risk for developing the disease (predictive testing). Testing should first be performed on an affected individual to determine whether or not there is an identifiable genetic or sporadic form of the disease in the family. Genetic testing identifies a mutation in a gene known to cause FTD approximately 10% of the time.

It is important to remember that genetic testing is voluntary. Because testing has both benefits and limitations, this decision is personal and complex. The physical risks associated with most genetic tests are small and involve a simple blood draw. Most of the risks associated with genetic testing involve possible emotional, social or financial consequences of the test results. You may feel sad, angry, frightened, depressed, anxious or guilty after learning your results. Learning that one carries a mutation in a gene that results in a progressive, lethal disorder can be devastating and should be considered very carefully.

On the other hand, test results can give you a sense of relief from uncertainty and help you make informed decisions about managing your health care. The test results might impact life decisions, such as career choice, family planning, or insurance coverage. Your genetic counselor can explain in detail the benefits, risks and limitations of a particular test. It is important that you understand and weigh these factors before making a decision.

If you decide to proceed with genetic testing, often a sample can often be collected following your consultation. The sample is then sent to a laboratory where technicians look for the specific changes in one of the four genes
associated with FTD. The laboratory will report the results to your doctor and/or genetic counselor who will discuss the results with you in person.

What do the results mean?

The results of genetic tests are not always straightforward, which often makes them challenging to interpret and explain. When interpreting test results, your doctor or genetic counselor considers your medical history, family history and the type of genetic test that was done.

A positive test result means that the laboratory detected a mutation (change) in a gene known to cause FTD. Depending on your health status, this result may confirm an existing diagnosis or it may indicate that you are at increased risk for developing FTD in the future. A positive test result may have implications for other blood relatives.

A negative test result means that the laboratory did not find a mutation in the tested gene. This negative test result must be interpreted with caution. Sometimes, a negative test result can suggest a sporadic form of the disease. In this case, other family members are not thought to be at increased risk for the condition. However, in other cases, a negative test result does not entirely exclude the possibility of a familial condition. It is important to discuss the test results with a genetic counselor or doctor to better understand the implications for yourself and family members.

In rare cases, the genetic test result is inconclusive. This occurs when the test identifies a natural variation in a gene, called a polymorphism. Polymorphisms are common in the population and, generally, do not affect your health. Sometimes it is difficult to know if a variation in a person's DNA is a very rare (uncommon) polymorphism or a mutation (a change in the DNA that will cause a genetic disease). If the genetic test reveals a change that has not previously been associated with a disorder in other people, this can sometimes be difficult to determine, and, therefore, results remain uncertain.

Genetic Information Nondiscrimination

The Genetic Information Nondiscrimination Act of 2008 (GINA) was enacted on May 21, 2008 to protect Americans against discrimination based on their genetic information when it comes to health insurance and employment. GINA prohibits group health plans and health insurers from denying coverage or charging higher premiums to a healthy individual based solely on a genetic predisposition to developing a disease in the future. The legislation also bars employers from using individuals’ genetic information when making hiring, firing or promotion decisions. This means that if you decide to undergo genetic testing, you may have to share your results with your employer or insurance company (most likely if they pay for the testing), but if the results are shared, they cannot be used to make decisions about your employment or health insurance coverage.

The insurance provisions of the bill took effect May 21, 2009 and the employment provisions went into effect on November 21, 2009.