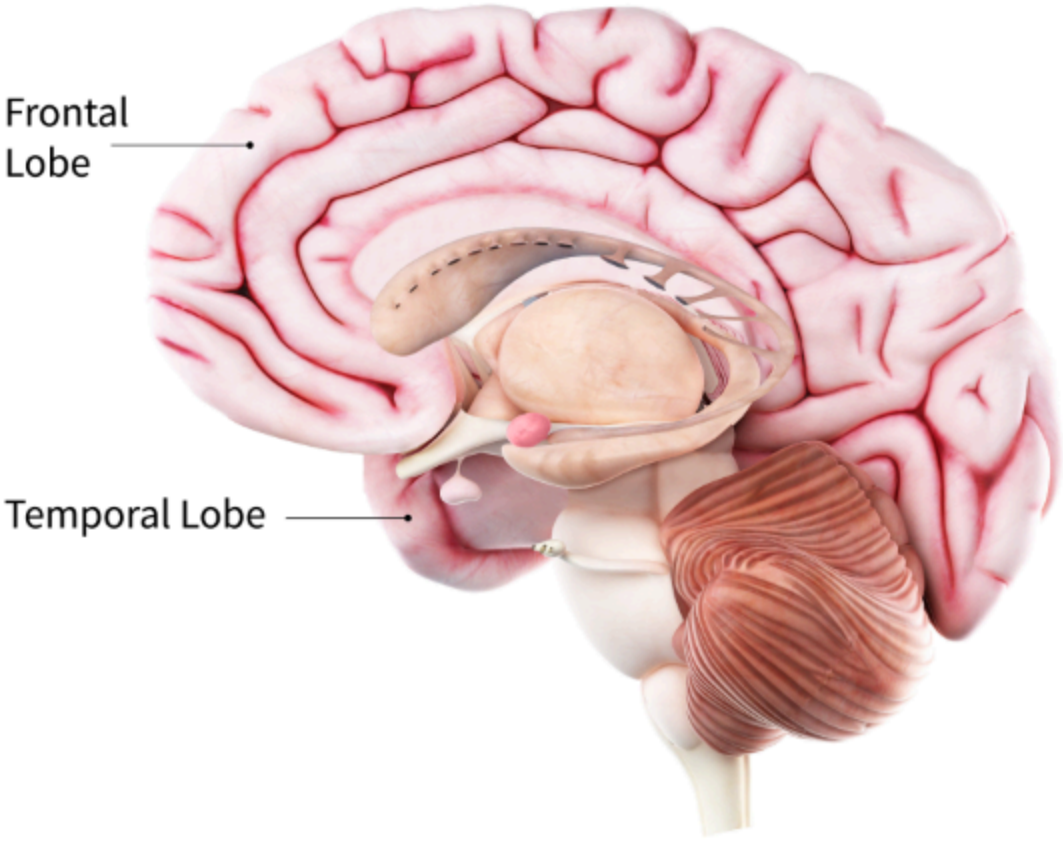
Share:

What Are Frontotemporal Disorders? Causes, Symptoms, and Treatment

On this page:

Frontotemporal disorders (FTD), sometimes called frontotemporal dementia, are the result of damage to neurons in the frontal and temporal lobes of the brain. Many possible symptoms can result, including unusual behaviors, emotional problems, trouble communicating, difficulty with work, or difficulty with walking. FTD is rare and tends to occur at a younger age than other forms of dementia. Roughly 60% of people with FTD are 45 to 64 years old.

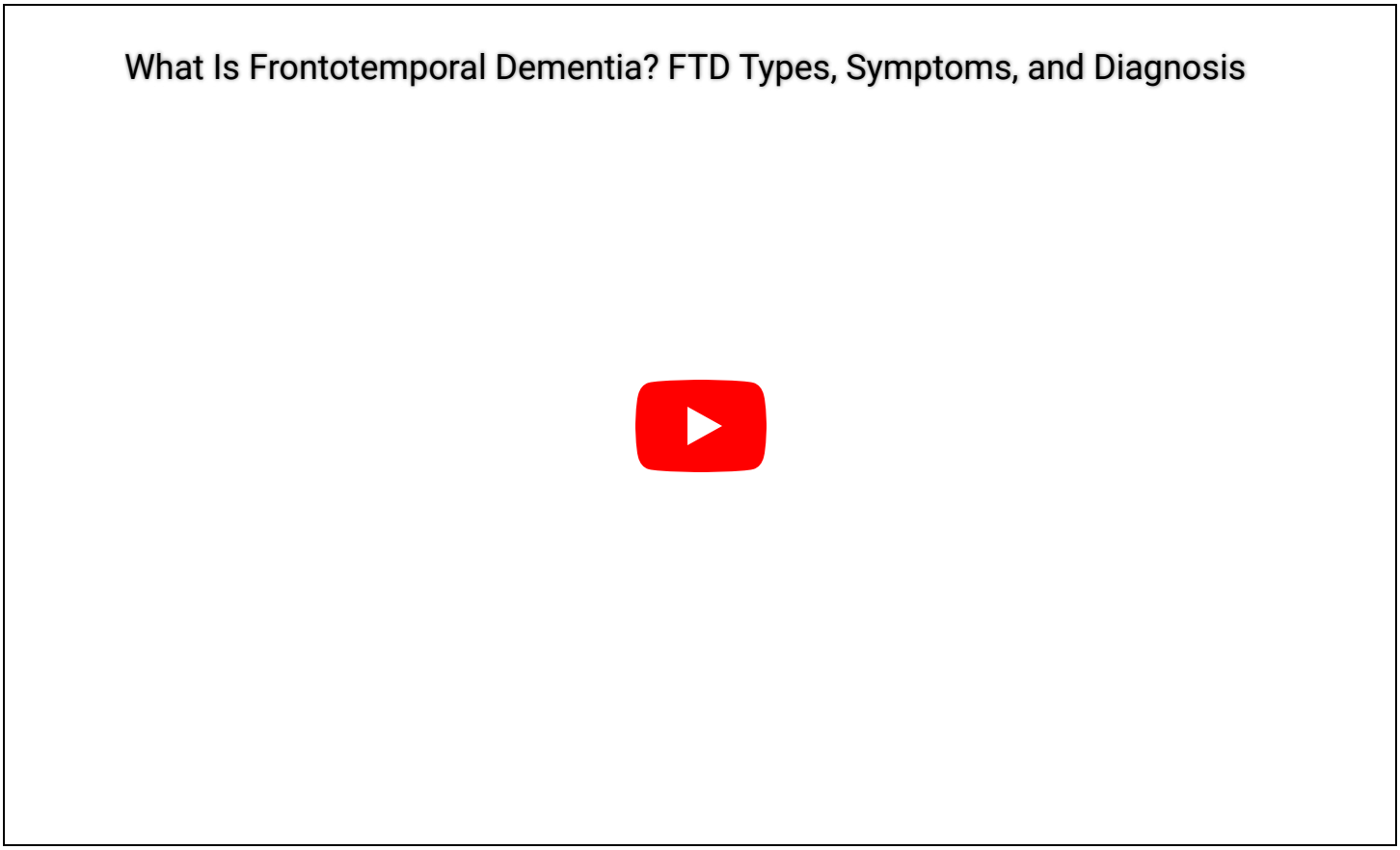


FTD is progressive, meaning symptoms get worse over time. In the early stages, people may have just one symptom. As the disease progresses, other symptoms appear as more parts of the brain are affected. It is difficult to predict how long someone with FTD will live. Some people live more than 10 years after diagnosis, while others live less than two years after they are diagnosed.

There is currently no cure for FTD, and no treatments slow or stop the progression of the disease, but there are ways to help manage the symptoms.

What do the terms mean?

One of the challenges shared by people living with these disorders, families, clinicians, and researchers is what terminology to use. Here, we have used the term frontotemporal disorders to characterize this group of diseases and the abbreviation FTD, which is commonly used to refer to them. Other terms used include frontotemporal lobar degeneration and frontotemporal dementia, but it's important to note that with some frontotemporal disorders, the primary symptoms are problems with speech or movement, rather than dementia symptoms. Physicians and psychologists diagnose the different forms of FTD based on a person's symptoms as well as the results of brain scans and genetic tests.



What are the types and symptoms of FTD?

In the early stages, it can be hard to know which type of FTD a person has because symptoms and the order in which they appear can vary from one person to another. Also, the same symptoms can appear across different disorders and vary from one stage of the disease to the next as different parts of the brain are affected.

Symptoms of FTD are often misunderstood. Family members and friends may think that a person is misbehaving, leading to anger and conflict. It is important to understand that people with these disorders cannot control their behaviors and other symptoms and lack any awareness of their illness.

There are three types of frontotemporal disorders (FTD): behavioral variant frontotemporal dementia (bvFTD), primary progressive aphasia (PPA), and movement disorders.

Behavioral variant frontotemporal dementia

The most common FTD, bvFTD, involves changes in personality, behavior, and judgment. People with this disorder may have problems with cognition, but their memory may stay relatively intact. Symptoms can include:

- Problems planning and sequencing (thinking through which steps come first, second, and so on)
- Difficulty prioritizing tasks or activities
- Repeating the same activity or saying the same word over and over
- Acting impulsively or saying or doing inappropriate things without considering how others perceive the behavior
- Becoming disinterested in family or activities they used to care about

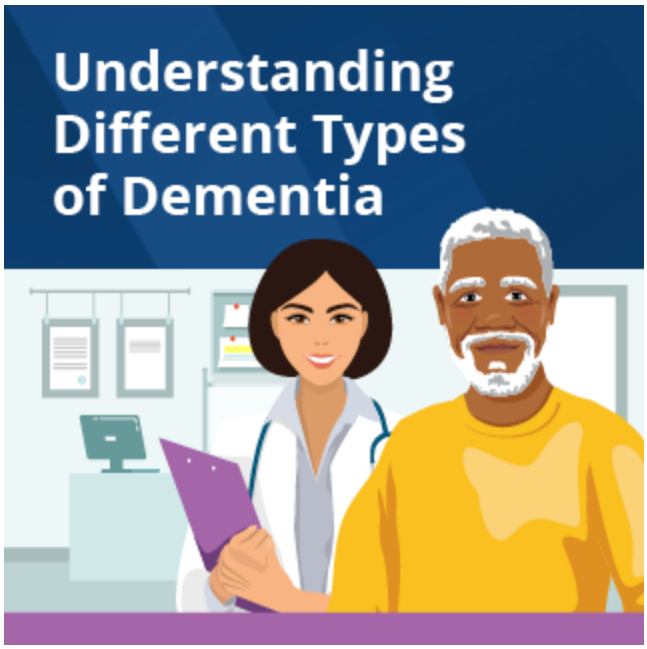
Over time, language and/or movement problems may occur, and the person living with bvFTD will need more care and supervision.

Primary progressive aphasia

PPA involves changes in the ability to communicate — to use language to speak, read, write, and understand what others are saying. This includes difficulty using or understanding words (aphasia) and difficulty speaking properly (e.g., slurred speech). People with PPA may have one or both of these symptoms. They may become mute or unable to speak.

Many people with PPA develop symptoms of dementia. Problems with memory, reasoning, and judgment are not apparent at first but can develop over time. In addition, some people with PPA may experience significant behavioral changes, similar to those seen in bvFTD, as the disease progresses.

There are three types of PPA, categorized by the kind of language problems that appear first.



Share this infographic and help spread the word about understanding different types of dementia.

- **Semantic PPA:** A person slowly loses the ability to understand single words and sometimes to recognize the faces of familiar people and common objects.
- **Agrammatic PPA:** A person has more and more trouble speaking and may omit words that link nouns and verbs (such as to, from, the). Eventually, the person may no longer be able to speak at all. The person may eventually develop movement symptoms similar to those seen in corticobasal syndrome.
- **Logopenic PPA:** A person has trouble finding the right words during a conversation but can understand words and sentences. The person does not have problems with grammar.

Researchers do not fully understand the biological basis of the different types of PPA. But they hope one day to link specific language problems with the changes in the brain that cause them.

Movement disorders

Two rare neurological movement disorders associated with FTD, corticobasal syndrome and progressive supranuclear palsy, occur when the parts of the brain that control movement are affected. The disorders may affect thinking and language abilities, too.

- **Corticobasal syndrome** can be caused by corticobasal degeneration — a gradual atrophy (shrinkage) and loss of nerve cells in specific parts of the brain. This degeneration causes progressive loss of the ability to control movement, typically beginning around age 60. The most prominent symptom may be apraxia, the inability to use the hands or arms to perform a movement despite normal strength, such as difficulty closing buttons or operating small appliances. Other symptoms can include muscle rigidity and difficulty swallowing. Symptoms may appear first on one side of the body, but eventually both sides are affected. Occasionally, a person with corticobasal syndrome first has language problems or trouble orienting objects in space and later develops movement symptoms. Not everyone who has corticobasal syndrome has problems with memory, cognition, language, or behavior.

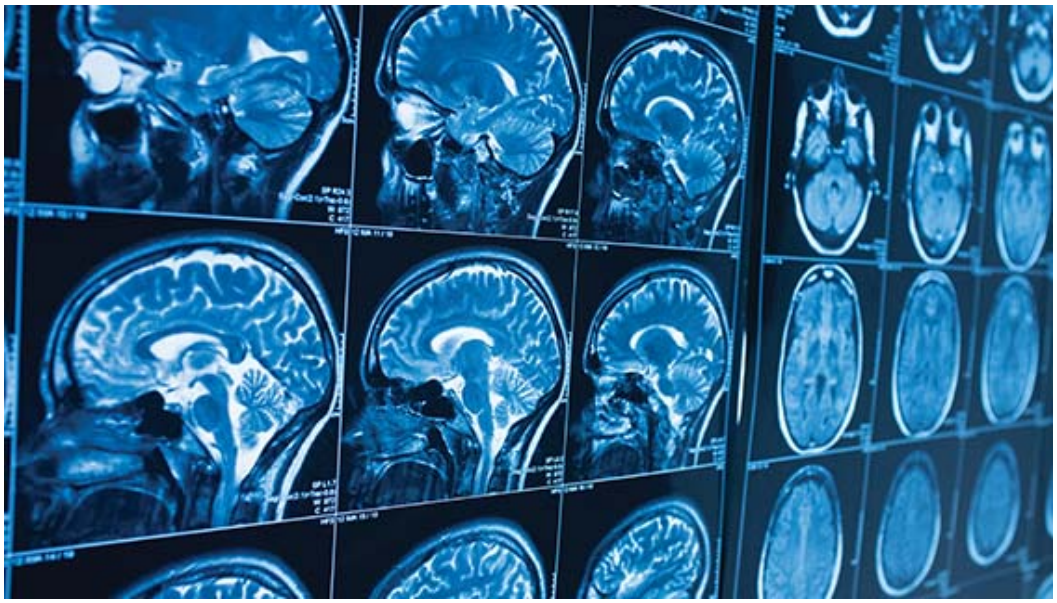
- **Progressive supranuclear palsy** causes problems with balance and walking. People with the disorder typically move slowly, experience unexplained falls, lose facial expression, and have body stiffness, especially in the neck and upper body — symptoms similar to those of Parkinson's disease. A hallmark sign of this disorder is trouble with eye movements, particularly looking down. These symptoms may give the face a fixed stare. Problems with behavior, memory, problem solving, and judgment can also develop.

Other movement-related types of FTD include frontotemporal dementia with parkinsonism and frontotemporal dementia with amyotrophic lateral sclerosis (FTD-ALS).

- **Frontotemporal dementia with parkinsonism** can be an inherited disease caused by a genetic tau genetic variant. Symptoms include movement problems similar to those of Parkinson's disease, such as slowed movement, stiffness, and balance problems, and changes in behavior or language.
- **FTD-ALS**, also called FTD with motor neuron disease, is a combination of bvFTD and ALS, the latter commonly known as Lou Gehrig's disease. In addition to the behavioral and/or language changes seen in bvFTD, people with FTD-ALS experience the progressive muscle weakness seen in ALS, fine jerks, and wiggling in muscles. Symptoms of either disease may appear first, with other symptoms developing over time. Changes in certain genes have been found in some people with FTD-ALS, though most cases are not hereditary.

What causes FTD?

Scientists are beginning to understand the biological and genetic basis for the changes observed in brain cells that lead to FTD.



Scientists describe FTD using the patterns of change in the brain seen in an autopsy after death. These changes include loss of neurons and abnormal amounts, or forms of proteins called tau and TDP-43. These proteins occur naturally in the body and help cells function properly. When the proteins don't work properly, for reasons not yet fully understood, neurons in specific brain regions are damaged.

In most cases, the cause of a FTD is unknown. Individuals with a family history of FTD are more likely to develop such a disorder. About 10 to 30% of bvFTD is due to specific genetic causes.

FTD that runs in a family is often related to variants (permanent changes) in certain genes. Genes are basic units of heredity that tell cells how to make the proteins the body needs to function. Even small changes in a gene may produce an abnormal protein, which can lead to changes in the brain and, eventually, disease.

Scientists have discovered several different genes that, when mutated, can lead to FTD:

- **Tau gene (also called the MAPT gene)** — A change in this gene causes abnormalities in a protein called tau, which then forms tangles inside neurons and ultimately leads to the destruction of brain cells. Inheriting a variant in this gene means a person will almost surely develop a frontotemporal disorder, usually bvFTD, but the exact age of onset and symptoms cannot be predicted.
- **GRN gene** — A change in this gene can lead to lower production of the protein progranulin, which in turn causes another protein, TDP-43, to go awry in brain cells. Many frontotemporal disorders can result, though bvFTD is the most common. The GRN gene can cause different symptoms in different family members and cause the disease to begin at different ages.
- **C9ORF72 gene** — An unusual change in this gene appears to be the most common genetic abnormality in familial frontotemporal disorders and familial ALS. This variant can cause a frontotemporal disorder, ALS, or both conditions.

In recent years researchers have discovered several other genetic changes in genes that lead to rare familial types of frontotemporal disorders. These other variants account for less than 5% of all cases of FTD.

Families affected by inherited and familial forms of FTD can help scientists advance research by participating in clinical studies and trials. For more information, talk with a health care professional or visit the Alzheimers.gov Clinical Trials Finder.

Brian's story

Brian, an attorney, began having trouble organizing his cases. In time, his law firm assigned him to do paperwork only. Brian's wife thought he was depressed because his father had died two years earlier. Brian, 56, was treated for depression, but his symptoms got worse. He became more disorganized and began making sexual comments to his wife's female friends. Even more unsettling, he neither understood nor cared that his behavior disturbed his family and friends. As time went on, Brian had trouble paying bills and was less affectionate toward his wife and young son. Three years after Brian's symptoms began, his counselor recommended a neurological evaluation. Brian was diagnosed with behavioral variant FTD — the most common form of FTD.

How is FTD diagnosed?

FTD can be hard to diagnose because the symptoms are similar to those of other conditions. For example, bvFTD is sometimes misdiagnosed as a mood disorder, such as depression. To make matters more confusing, a person can have both FTD and another type of dementia, such as Alzheimer's disease. Also, because these disorders are rare, physicians may be unfamiliar with the signs and symptoms.

To help diagnose frontotemporal dementia, a doctor may:

- Perform an exam and ask about symptoms
- Look at personal and family medical history
- Use laboratory tests to help rule out other conditions
- Order genetic testing
- Conduct tests to assess memory, thinking, language skills, and physical functioning
- Order imaging of the brain

A psychiatric evaluation can help determine if depression or another mental health condition is causing or contributing to the condition. Only genetic tests in familial cases or a brain autopsy after a person dies can confirm a diagnosis of FTD.

Researchers are studying ways to diagnose FTD earlier and more accurately and to distinguish them from other types of dementia. One area of research involves biomarkers, such as proteins or other substances in the blood or cerebrospinal fluid which can be used to measure disease progression or the effects of treatment. Researchers are also exploring ways to improve brain imaging and neuropsychological testing.

Treatment and management of FTD

So far, there is no cure for FTD and no way to slow down or prevent these diseases. However, there are ways to manage symptoms. A team of specialists — doctors, nurses, and speech, physical, and occupational therapists — familiar with these disorders can help guide treatment.

Clinical trials on frontotemporal disorders

> Volunteers are needed for clinical trials that are testing treatments for FTD. By joining one of these studies, you may learn more about how to manage FTD symptoms and contribute useful information to help others in the future.

Managing behavior changes in FTD

Behavior changes associated with bvFTD can upset and frustrate family members and other caregivers. Understanding changes in personality and behavior and knowing how to respond can reduce frustration and help provide the best care for a person with FTD.

Managing behavioral symptoms can involve several approaches. Here are some strategies to consider:

- Try to accept rather than challenge someone with behavioral symptoms. Arguing or reasoning will not help, because they cannot control their behaviors or see that they are unusual or upsetting to others. Instead, be as sensitive as possible and understand that it's the illness "talking."
- Take a "timeout" when frustrated — take deep breaths, count to 10, or leave the room for a few minutes.
- To deal with apathy, limit choices and offer specific choices. Open-ended questions, such as "What do you want to do today?" are more difficult to answer than specific ones, such as "Do you want to go to the park or for a walk?".
- Maintain a regular schedule, reduce distractions, and modify the environment to reduce confusion and improve the person's sleep.
- If compulsive eating is an issue, consider supervising eating, limiting food choices, locking cabinets and the refrigerator, and distracting the person with other activities.

To ensure the safety of a person and his or her family, caregivers may have to take on new responsibilities or arrange care that was not needed before.

Medications are available to treat certain behavioral symptoms. Antidepressants called selective serotonin reuptake inhibitors are commonly prescribed to treat social disinhibition and impulsive behavior. People with aggression or delusions sometimes take low doses of antipsychotic medications. If a particular medication is not working, a doctor may try another. Always consult a doctor before changing, adding, or stopping a drug or supplement.

Treating language problems in FTD

Treatment of PPA has two goals — maintaining language skills and using new tools and other ways to communicate. Treatment tailored to a person's specific language problem and stage of PPA generally works best. Since language ability declines over time, different strategies may be needed as the illness progresses. The following strategies may help:

- Use a communication notebook (an album of photos labeled with names of people and objects), gestures, and drawings to communicate without talking.
- Store lists of words or phrases in a computer or phone to point to.
- Speak slowly and clearly, use simple sentences, wait for responses, and ask for clarification if needed.

- Work with a speech-language pathologist familiar with PPA to determine the best tools and strategies to use. Note that many speech-language pathologists are trained to treat aphasia caused by stroke, which requires different strategies from those used with PPA.

> **Mary Ann's story**
>
> Mary Ann, a television news anchor for 20 years, began having trouble reading the nightly news. At first, her doctor thought she had a vision problem, but tests showed that her eyesight was normal. Although normally creative and energetic, Mary Ann, 52, had trouble finishing assignments and voicing her ideas at staff meetings. In time, she was let go from her job. Mary Ann applied for Social Security disability benefits, which required a medical exam. Her symptoms puzzled several doctors until a neurologist diagnosed logopenic PPA. A speech therapist taught Mary Ann to use a personal digital assistant to express words and phrases. For emergencies, Mary Ann carries a card in her wallet that explains her condition.

Managing movement problems in FTD

Medications and physical and occupational therapy may provide modest relief for the movement symptoms of FTD. A doctor who specializes in these disorders can guide treatment.

For people with corticobasal syndrome, Parkinson's disease medicines may offer some temporary improvement. Physical and occupational therapy may help the person move more easily. Speech therapy can help them manage language symptoms.

For people with progressive supranuclear palsy, sometimes Parkinson's disease drugs provide temporary relief for slowness, stiffness, and balance problems. Exercises can keep the joints limber, and weighted walking aids — such as a walker with sandbags over the lower front rung — can help maintain balance. Speech, vision, and swallowing difficulties usually do not respond to any drug treatment. Antidepressants have shown modest success. For people with abnormal eye movements, bifocals or special glasses called prisms are sometimes prescribed.

People with FTD-ALS typically decline quickly over two to three years. During this time, physical therapy can help treat muscle symptoms, and a walker or wheelchair may be useful. Speech therapy may help a person speak more

clearly at first. Later on, other ways of communicating, such as a speech synthesizer, can be used. The ALS symptoms of the disorder ultimately make it impossible to stand, walk, eat, and breathe on one's own.

Physicians, nurses, social workers, and physical, occupational, and speech therapists who are familiar with these conditions can ensure that people with movement disorders get appropriate medical treatment and that their caregivers can help them live as well as possible.

The future of FTD treatment

Researchers are continuing to explore the biological changes in the body, including genetic variants and proteins, that lead to FTD and identify and test possible new drugs and other treatments. They are also developing better ways to track disease progression, so that treatments, when they become available, can be directed to the right people. Clinical trials and studies are underway to advance these efforts. People with FTD and healthy people may be able to participate. To find out more, talk to your health care provider or visit the Alzheimers.gov Clinical Trials Finder.

Where to find FTD diagnosis and treatment

Columbia-Presbyterian Medical Center
Department of Neurology
New York, NY
646-426-3876

Houston Methodist Hospital
Frontotemporal Degeneration Unit
Houston, TX
713-441-7650

Indiana University School of Medicine
Indiana Alzheimer's Disease Center
Indianapolis, IN
317-963-5500

Johns Hopkins University School of Medicine
Frontotemporal Dementia and Young-Onset Dementias Clinic
Baltimore, MD
410-955-5147

Massachusetts General Hospital
Frontotemporal Disorders Unit
Boston, MA
617-726-1728

Mayo Clinic
Department of Neurology
Rochester, MN
507-538-3270
Jacksonville, FL
904-953-0853
Phoenix or Scottsdale, AZ
800-446-2279

Northwestern University Feinberg School of Medicine
Mesulam Center for Cognitive Neurology and Alzheimer's Disease
Chicago, IL
312-908-9339

University of Alabama, Birmingham
Neurology Department, Division of Memory Disorders
Birmingham, AL
205-996-3679

University of California, Los Angeles
Neurobehavior Clinic
Los Angeles, CA
310-794-1195

University of California, San Diego
Shiley-Marcos Alzheimer's Disease Research Center
La Jolla, CA
858-822-4800

University of California, San Francisco
Memory and Aging Center
San Francisco, CA
415-353-2057

University of Pennsylvania Health System
Penn Frontotemporal Degeneration Center
Philadelphia, PA
215-349-5863

Washington University
Department of Neurology
St. Louis, MO
314-362-1408

Subscribe

For more information about FTD

NIA Alzheimer's and related Dementias Education and Referral (ADEAR) Center
800-438-4380
adear@nia.nih.gov
www.nia.nih.gov/alzheimers
The NIA ADEAR Center offers information and free print publications about Alzheimer's and related dementias for families, caregivers, and health professionals. ADEAR Center staff answer telephone, email, and written requests and make referrals to local and national resources.

Alzheimers.gov
www.alzheimers.gov
Explore the Alzheimers.gov website for information and resources on Alzheimer's and related dementias from across the federal government.

Association for Frontotemporal Degeneration
866-507-7222
info@theaftd.org
www.theaftd.org

National Institute of Neurological Disorders and Stroke (NINDS)
800-352-9424
braininfo@ninds.nih.gov
www.ninds.nih.gov

MedlinePlus
National Library of Medicine
www.medlineplus.gov

Eldercare Locator
800-677-1116
eldercarelocator@USAging.org
https://eldercare.acl.gov

This content is provided by the NIH National Institute on Aging (NIA). NIA scientists and other experts review this content to ensure it is accurate and up to date.

Content reviewed: July 30, 2021

Return to top

Newsletters

Sign up to receive updates and resources delivered to your inbox.

Sign up