

**FRONTOTEMPORAL
DISORDERS:
INFORMATION FOR
PATIENTS,....**

**NATIONAL
INSTITUTE OF
AGING**



FRONTOTEMPORAL DISORDERS

Information for Patients, Families,
and Caregivers

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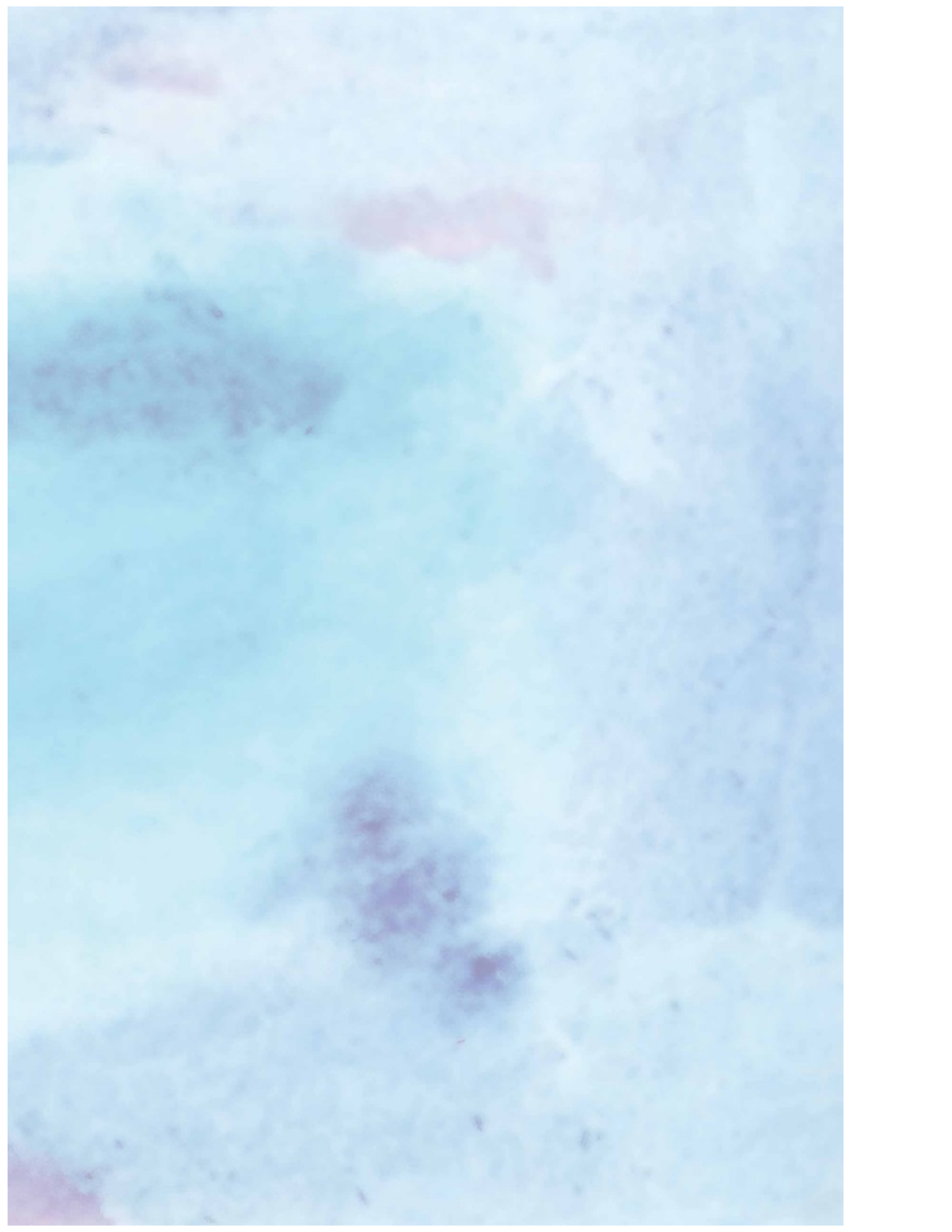
- F
- frontotemporal dementia
- P
- primary progressive aphasia
- Movement disorders
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The National Institute on Aging (NIA), part of the National Institutes of Health, leads the Federal Government effort conducting and supporting research on Alzheimer's disease and other forms of dementia. NIA's Alzheimer's Disease Education and Referral (ADEAR) Center offers information and publications on dementia and caregiving for families, caregivers, and professionals. For more information, visit www.nia.nih.gov/alzheimers or call **1-800-438-4380**.



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Introduction

Few people have heard of frontotemporal dementia and other brain disorders that affect personality, behavior, language, and movement.

These disorders are little known outside the circles of researchers, doctors, patients, and caregivers who study and live with them. Although frontotemporal disorders remain puzzling in many ways, researchers are finding new clues that will help them solve this medical mystery and better understand other common dementias.

The symptoms of frontotemporal disorders are devastating. They gradually rob people of basic abilities—thinking, talking, walking, and socializing—

that most of us take for granted. They often strike people in the prime of life, when they are working and raising families. Families suffer, too, as they struggle to cope with the person's daily needs as well as changes in relationships and finances.

This booklet is meant to help people with frontotemporal disorders, their families, and caregivers learn more about these conditions and resources for coping. It explains what is known about the different types of disorders and how they are diagnosed. Most importantly, it describes how to treat and manage these difficult conditions, with practical advice for caregivers. A list of resources begins on page 27.

1



The Basics of Frontotemporal Disorders

frontal lobe

temporal

Frontotemporal disorders are the result

of damage to neurons (nerve cells)

lobe

in parts of the brain called the frontal

and temporal lobes. As neurons die in the

frontal and temporal regions, these lobes

atrophy, or shrink. Gradually, this damage

causes difficulties in thinking and behaviors

controlled by these parts of the brain. Many

possible symptoms can result, including strange behaviors, emotional problems, trouble communicating, or difficulty with walking and other basic movements.

A Form of Dementia

Frontotemporal disorders are a form of dementia caused by a family of brain diseases known as frontotemporal lobar degeneration (FTLD). Dementia is a severe loss of thinking abilities that interferes with a person's ability to perform daily activities such as working, driving, and preparing meals.

Other brain diseases that can cause dementia include Alzheimer's disease and strokes. Scientists estimate that FTLD may cause up to 10 percent of all cases of dementia and may be about as common as Alzheimer's among people younger than age 65.

People can live with frontotemporal disorders for 2 to 10 years, sometimes longer, but it is difficult to predict the time course for an individual patient.

The disorders are progressive, meaning symptoms get worse over time. In the early stages, people have one type of symptom. As the disease progresses, other types of symptoms appear as more parts of the brain are affected.

No cure or treatments for frontotemporal disorders are available today.

However, research is improving awareness and understanding of these challenging conditions. This progress is opening doors to better diagnosis, improved care, and, eventually, possible new treatments.





FTD? FTLD? Understanding Terms

One of the challenges shared by patients, families, clinicians, and researchers is confusion about how to classify and label frontotemporal disorders. A diagnosis by one doctor may be called something else by a second, and the same condition or syndrome referred to by another name by a pathologist who examines the brain after death.

For many years, scientists and physicians used the term *frontotemporal dementia* (FTD) to describe this group of illnesses. After further research, FTD is now understood to be just one of several possible variations and is more precisely called *behavioral variant frontotemporal dementia*, or bvFTD.

This booklet uses the term *frontotemporal disorders* to refer to changes in behavior and thinking that are caused by underlying brain diseases collectively called *frontotemporal lobar degeneration* (FTLD). FTLD is not a single brain disease but rather a family of neurodegenerative diseases, any one of which can cause a frontotemporal disorder (see “Causes,” page 11).

Frontotemporal disorders are diagnosed by physicians and psychologists based on a person's symptoms. FTLD can be identified definitively only by brain autopsy after death.

Changes in the Brain

Frontotemporal disorders affect the frontal and temporal lobes of the brain.

They can begin in the frontal lobe, the temporal lobe, or both. Initially, frontotemporal disorders leave other brain regions untouched, including those that control short-term memory.

The frontal lobes, situated above the eyes and behind the forehead both on the right and left sides of the brain, direct executive functioning. This includes planning and sequencing (thinking through which steps come first, second, third, and so on), prioritizing (doing more important activities first and less important activities last), multitasking (shifting from one activity to another as needed), and monitoring and correcting errors.

3





What's going on?

Brian, an attorney, began having trouble organizing his cases .

When functioning well, the In time, his law firm frontal lobes also help manage assigned him to do paperwork only . Brian's emotional responses. They enable wife thought he was depressed because people to control inappropriate his father had died 2 years earlier . Brian, social behaviors, such as shouting 56, was treated for depression, but his loudly in a library or at a funeral. symptoms got worse . He became more disorganized and began making sexual

They help people make decisions comments to his wife's female friends .

that make sense for a given situ-

Even more unsettling, he neither under-

ation. When the frontal lobes are

stood nor cared that his behavior

disturbed his family and friends . As time

damaged, people may focus on

went on, Brian had trouble paying bills

insignificant details and ignore

and was less affectionate toward his

important aspects of a situation

wife and young son . Three years after

Brian's symptoms began, his counselor

or engage in purposeless activities.

recommended a neurological evaluation .

The frontal lobes are also involved

Brian was diagnosed with bvFTD .

in language, particularly linking

words to form sentences, and in

motor functions, such as moving the arms, legs, and mouth.

The temporal lobes, located below and to the side of each frontal lobe on the right and left sides of the brain, play a major role in language and emotions. They help people understand words, speak, read, write, and connect words with their meanings. They allow people to recognize objects, including faces, and to relate appropriate emotions to objects and events.

When temporal lobes are dysfunctional, people may have difficulty recognizing emotions and responding appropriately to them.

Which lobe—and part of the lobe—is affected first determines which symptoms appear first. For example, if the disease starts in the part of the frontal lobe responsible for decision-making, then the first symptom might be trouble managing finances. If it begins in the part of the temporal lobe that connects emotions to objects, then the first symptom might be an inability to recognize potentially

dangerous objects—a person might reach for a snake or plunge a hand into boiling water, for example.

4





Types of Frontotemporal Disorders

Frontotemporal disorders can be grouped into three types, defined by the earliest symptoms physicians identify when they examine patients.

- **Progressive behavior/personality decline**—characterized by changes in personality, behavior, emotions, and judgment (e.g., behavioral variant frontotemporal dementia).
- **Progressive language decline**—marked by early changes in language ability, including speaking, understanding, reading, and writing (e.g., primary progressive aphasia).
- **Progressive motor decline**—characterized by various difficulties with physical movement, including shaking, difficulty walking, frequent falls, and poor coordination.

It can be hard to know which of these disorders a person has because symptoms and the order in which they appear can vary widely from one person to the next. Also, the same

symptoms can appear in different disorders. For example, language

Trouble with words

problems are most typical of

Alicia's first symptom

primary progressive aphasia but

was trouble talking .

can also appear in the course of

She spoke more

slowly and thought

behavioral variant frontotemporal

she sounded stilted .

dementia. The table on page 6

She could under-

summarizes the three types of

stand people well enough, but finding

the right words when she was talking

frontotemporal disorders and

became harder and harder . Also, Alicia,

lists the various terms that could

49, could not write words like “and”

be used when clinicians diagnose

and “it” but could write words like

“alligator .” Her doctor recommended

these disorders.

a neurological exam, which helped

diagnose agrammatic PPA .

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Types of Frontotemporal Disorders

Diagnostic Terms

Main Early Symptoms

Progressive Behavior/Personality Decline

- Behavioral variant
- Apathy, reduced initiative

frontotemporal dementia

- Inappropriate and impulsive behaviors

(bvFTD)

- Emotional flatness or excessive emotions
- Temporal/frontal variant FTD • Memory generally intact (tvFTD, fvFTD)
- Pick's disease

Progressive Language Decline

- Primary progressive aphasia • Semantic PPA (also called semantic dementia): (PPA)

can't understand words or recognize familiar

- Progressive nonfluent

people and objects

aphasia

- Agrammatic PPA (also called progressive nonfluent aphasia): omits words that link nouns and verbs (such as to, from, the); difficulty swallowing
- Semantic dementia

nonfluent aphasia): omits words that link

nouns and verbs (such as to, from, the);

difficulty swallowing

- Logopenic PPA: trouble finding the right

words while speaking, hesitation, and/or

pauses in speech

Progressive Motor Decline

- Corticobasal syndrome
- Muscle rigidity

(CBS)

- Difficulty closing buttons, operating simple appliances

- Language or spatial orientation problems

- Progressive supranuclear

- Progressive problems with balance and

palsy (PSP)

walking

- Slow movement, falling, body stiffness

- Restricted eye movements

- FTD with parkinsonism

- Movement problems similar to Parkinson's

disease, such as slowed movement and

stiffness

- Changes in behavior or language

- FTD with amyotrophic lateral • Combination of FTD and ALS (Lou Gehrig's sclerosis (FTD-ALS) disease)

- Changes in behavior and/or language

- Muscle weakness, shrinkage, jerking





Behavioral Variant Frontotemporal Dementia

The most common frontotemporal disorder, *behavioral variant frontotemporal dementia* (bvFTD), involves changes in personality, behavior, and judgment.

People with this dementia can act strangely around other people, resulting in embarrassing social situations. Often, they don't know or care that their behavior is unusual and don't show any consideration for the feelings of others. Over time, language and/or movement problems may occur, and the person needs more care and supervision.

In the past, bvFTD was called Pick's disease, named after Arnold Pick, the German scientist who first described it in 1892. The term Pick's disease is now used to describe abnormal collections in the brain of the protein tau, called "Pick bodies," which can only be seen under the microscope after death. Some patients with bvFTD have Pick's disease, and some do not.

Primary Progressive Aphasia

Primary progressive aphasia (PPA) involves changes in the ability to communicate—to use language to speak, read, write, and understand what others are saying. Problems with memory, reasoning, and judgment are not apparent at first but can develop over time. In addition, some people with 7





“What do you mean by salt?”

Jane, 62, a university professor, began having trouble remembering the names of common objects while she lectured . She also had a hard time following conversations, especially when more than one person was involved . Her family and co-workers were unaware of Jane’s difficulties—until she had a hard time recognizing . Currently, there are three types of

longtime colleagues . One night at the

PPA, categorized by the kind of

dinner table, when Jane’s husband

language problems seen at first.

asked her to pass the salt, she said,

Researchers do not fully under-

“Salt? What do you mean by salt?”

He took her to a neurologist, who

stand the different types of PPA.

diagnosed semantic PPA . As her illness

But they hope one day to link

progressed, Jane developed behavioral

specific language problems with

symptoms and had to retire early .

the abnormalities in the brain

that cause them.

In *semantic PPA*, also called semantic dementia, a person slowly loses the ability to understand single words and sometimes to recognize the faces of familiar people and common objects.

In *agrammatic PPA*, also called progressive nonfluent aphasia, a person has trouble saying words that link nouns and verbs together—for example, “of,”

“from,” and “for.” Eventually, the person may no longer be able to speak at all. He or she may also have difficulty swallowing and develop movement symptoms similar to those seen in corticobasal syndrome.

In *logopenic PPA*, a person has trouble finding the right words during conversation but can understand words and sentences. The person does not have problems with grammar.



Movement Disorders

Two rare neurological disorders associated with FTLN, *corticobasal syndrome* (CBS) and *progressive*

supranuclear palsy (PSP), occur when the nerves attached to muscles malfunction and cause problems with movement.

The disorders may affect thinking and language abilities, too.

CBS is caused by corticobasal degeneration—gradual atrophy and loss of nerve cells in specific parts of the brain. This degeneration causes progressive loss of movement and muscle rigidity, typically beginning around age 60.

Symptoms may appear first on one side of the body, but eventually both sides are affected. Occasionally, a person with CBS first has language problems or trouble orienting objects in space and later develops movement symptoms.

PSP causes serious problems with balance and walking. People with the disorder typically move slowly, fall, and have body stiffness, especially in the neck and upper body—symptoms similar to those of Parkinson’s disease. A hallmark sign of PSP is trouble with eye movements, such as involuntary closing of the eyelids,

difficulty opening and closing the eyes, trouble looking way up and

Confusing symptoms

way down, and limited blinking.

Carol had a tingling

These symptoms may give the face

sensation and numb-

a fixed stare. Behavior problems

ness in her upper

can also develop.

right arm . Then her

arm became stiff . She

had to change from

Other movement-related fron-

cursive handwriting to printing . Carol,

temporal disorders include

61, told her doctor that she had trouble
frontotemporal dementia with
getting her thoughts out and described
her speech as “stumbling .” She had
parkinsonism and frontotemporal
increasing trouble talking but could still
dementia with amyotrophic lateral
understand others . Eventually, she was
sclerosis (FTD-ALS).
diagnosed with CBS .

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Trouble with walking

For a year and a half, John had trouble walking and Frontotemporal dementia with fell several times . parkinsonism is an inherited He also had trouble disease caused by a genetic concentrating . He couldn't read because the words mutation. Symptoms include merged together on the page . John, movement problems similar to 73, also seemed less interested in those of Parkinson's disease, such social activities and projects around the house . His wife noticed that he as slowed movement, stiffness, and was more irritable than usual and balance problems, and changes in sometimes said uncharacteristically behavior or language. inappropriate things . John's primary care doctor did several tests, then FTD-ALS is a combination of

referred him to a neurologist, who

diagnosed PSP .

bvFTD and ALS, commonly

called Lou Gehrig's disease.

Symptoms include the behavioral

and/or language changes seen in bvFTD as well as the muscle weakness, shrinking, and jerking seen in ALS. Symptoms of either disease may appear first, with other symptoms developing over time.

Mutations in certain genes have been found in some patients with FTD-ALS.

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Causes

Frontotemporal lobar degeneration (FTLD) is not a single brain disease but rather a family of brain diseases that share some common molecular features. Scientists are just beginning to understand the biological and genetic basis for the changes observed in brain cells that lead to FTLD.

Scientists describe FTLD in terms of physical changes 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 the frontal and/or temporal lobes are damaged and disease results.

About 20 to 40 percent of people with frontotemporal disorders have a family history of them. About 10 percent of people inherit them directly from a parent. In most cases, the cause is unknown.

Familial and inherited forms of frontotemporal disorders are often related to mutations (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 frontotemporal disorders:

- **Tau gene (also called the MAPT gene)**—A mutation in this gene causes abnormal tau to form, which leads to the destruction of brain cells. Inheriting a mutation 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.



- **PGRN gene**—A mutation in this gene can lead to lower production of the protein progranulin, which in turn causes TDP-43, a cellular protein, to go awry. The result is familial bvFTD and possibly a higher chance of developing PPA.
- **VCP gene and CHMP2B gene**—Mutations in these genes lead to very rare familial types of frontotemporal disorders.
- **C9ORF72 gene**—An unusual mutation in this gene appears to be the most common genetic abnormality in familial frontotemporal disorders.

It is also the most common genetic abnormality in familial ALS and occurs in some cases of sporadic ALS.

Scientists are continuing to search for other genes and proteins that may play a role in frontotemporal disorders. For example, some researchers are exploring the FUS gene, which causes a type of familial ALS. Scientists are also trying to understand how mutations in a single gene lead to different frontotemporal disorders in different family members.



Diagnosis

No single test, such as a blood test, can be used to diagnose a frontotemporal disorder. A definitive diagnosis can be confirmed only by a brain autopsy after a person dies. To diagnose a probable frontotemporal disorder in a living person, a doctor—usually a neurologist, psychiatrist, or psychologist—will:

- record a person's symptoms, often with the help of family members or friends
- compile a personal and family medical history
- perform a physical exam and order blood tests to help rule out other similar conditions
- conduct a neuropsychological evaluation to assess behavior, language, memory, and other cognitive functions
- use brain imaging to look for changes in the frontal and temporal lobes.

Different types of brain imaging

may be used. A magnetic

Is it depression?

resonance imaging (MRI) scan

shows changes in the size and

Ana's husband was

the first to notice

shape of the brain, including the

a change in his

frontal and temporal lobes. It

55-year-old wife's

may reveal other causes of the

personality . Normally

active in her commu-

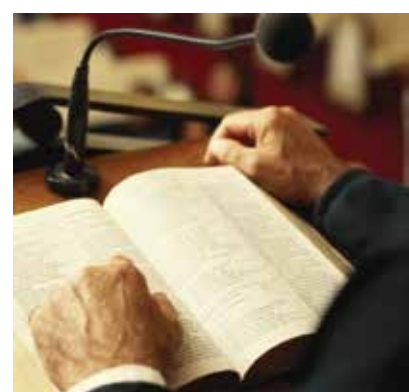
person's symptoms, such as a

nity, she became less interested in her

stroke or tumor. In the early stage
volunteer activities . She wanted to stay
of disease, the MRI result may
home, did not initiate conversations,
and went on her daily walks only if her
appear normal. In this case, other
husband suggested it . Ana's family
types of imaging, such as positron
thought she might be depressed . A
emission tomography (PET) or
psychologist referred her to a neuro-
logist, who diagnosed bvFTD .
single photon emission computed
tomography (SPECT), may be

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Can't find the right words

Ray, 60, was a small-town minister, a quiet intellectual, and a

useful. PET and SPECT scans

poet . He had always

measure activity in the brain by

been thoughtful and

monitoring blood flow, glucose

well spoken, but

over time his weekly sermons became

usage, and other processes.

halting and confused . He had trouble

finding the right words and resorted to

Frontotemporal disorders can

gestures . His family doctor thought that

be hard to diagnose because

Ray had had a stroke . But when tests

their symptoms—changes in

showed no stroke, she referred Ray to

a university medical center, where

personality and behavior and dif-

specialists diagnosed logopenic PPA .

ficulties with speech and move-

ment—are similar to those of

other conditions. For example,

bvFTD is sometimes misdiagnosed as a mood disorder, such as depression, or as a stroke, especially when there are speech or movement problems. To make matters more confusing, a person can have both a frontotemporal disorder and another type of dementia. Also, since these disorders are rare, physicians may be unfamiliar with the relevant symptoms and signs.

Getting the wrong diagnosis can be frustrating. Without knowing their true condition, people with

frontotemporal disorders may not get appropriate treatment to manage their symptoms. Families may not get the help they need. People lose valuable time needed to plan treatment and future care. The medical centers listed on pages 29–30 of this booklet are places where people with frontotemporal disorders can be diagnosed and treated.

Researchers are studying ways to diagnose frontotemporal disorders earlier and more accurately. One area of research involves biomarkers, such as proteins or other substances in the blood or cerebrospinal fluid, which can be used to measure the progress of disease or the effects of treatment. Also being studied are ways to improve brain imaging and neuropsychological testing, which assesses learning, language, problem solving, memory, and other thinking skills.

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Common Symptoms

Symptoms of frontotemporal disorders vary from person to person and from one stage of the disease to the next as different parts of the frontal and temporal lobes are affected. In general, changes in the frontal lobe are associated with behavioral symptoms, while changes in the temporal lobe lead to language and emotional disorders.

Symptoms are often misunderstood. Family members and friends may think that a person is misbehaving, leading to anger and conflict. For example, a person with bvFTD may neglect personal hygiene or start shoplifting. It is important to understand that people with these disorders cannot control their behaviors and other symptoms.

Behavioral Symptoms

- **Problems with executive functioning**—Problems with planning and sequencing (thinking through which steps come first, second, third, and so on), prioritizing (doing more important activities first and less important activities last), multitasking (shifting from one activity to another as needed), and self-monitoring and correcting behavior.
- **Perseveration**—A tendency to repeat the same activity or to say the same word over and over, even when it no longer makes sense.
- **Social disinhibition**—Acting impulsively without considering how others perceive the behavior. For example, a person might hum at a business meeting or laugh at a funeral.
- **Compulsive eating**—Gorging on food, especially starchy foods like bread and cookies. People may take food from other people's plates.
- **Utilization behavior**—Difficulty resisting impulses to use or touch objects that one can see and reach. For example, a person picks up the telephone receiver while walking past it when the phone is not

ringing and the person does not intend to place a call.

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Language Symptoms

- **Aphasia**—A language disorder in which the ability to use or understand words is impaired but the physical ability to speak properly is normal.
- **Dysarthria**—A language disorder in which the physical ability to speak properly is impaired (e.g., slurring) but the message is normal.

People with PPA may have only problems using and understanding words or also problems with the physical ability to speak. People with both kinds of problems have trouble speaking and writing and may become mute, or unable to speak. Language problems usually get worse for at least 2 years after they appear. Other thinking and social skills remain normal longer before deteriorating.

Emotional Symptoms

- **Apathy**

Embarrassing behavior

—A lack of interest,

David and his wife

drive, or initiative. Apathy is

ran a successful store

often confused with depression,

until he began to act

but people with apathy may

strangely . He intruded

on his teenaged

not be sad. They often have

daughters' gatherings

trouble starting activities but
with friends, standing and staring at
can participate if others do
them but not realizing how embarrassed
the planning.

they were . He took food from other
people's plates . A year later, David, 47,

• **Emotional changes—**

and his wife lost their business . After

Emotions are flat, exaggerated,

a misdiagnosis of depression and no

improvement, David's wife took him to a

or improper. Emotions may

neurologist, who diagnosed bvFTD .

seem completely disconnected

from a situation or are

expressed at the wrong times

or in the wrong circumstances.

For example, a person may

laugh at sad news.





Not acting like himself

Previously a devoted
husband, Gary began

• **Social-interpersonal**

an extramarital affair

changes—Difficulty “reading”

at age 55—and

social signals, such as facial

didn’t care that every-

one knew about it .

expressions, and understanding

His wife was devastated and angry .

personal relationships. People

His friends agreed that this was not like

may lack empathy—the ability

him . All attempts to reason with him

were unsuccessful, as Gary could not

to understand how others are

understand how his actions hurt others .

feeling—making them seem

His wife insisted on a visit to the doctor .

indifferent, uncaring, or selfish.

Initially, Gary was misdiagnosed with

bipolar disorder . After further evaluation,

For example, the person may

he was told he had bvFTD .

show no emotional reaction to

illnesses or accidents that occur

to family members.

- **Changes in insight**—An inability to understand changes in one’s condition and how they affect others. This lack of awareness is hard for caregivers to deal with because the person may reject efforts to help.

Movement Symptoms

- **Dystonia**—Abnormal postures of the hands or feet. A limb may be bent stiffly or not used when performing activities that are normally done with two hands.
- **Gait disorder**—Abnormalities in walking, such as walking with a shuffle, sometimes with frequent falls.
- **Tremor**—Shakiness, usually of the hands.
- **Clumsiness**—Dropping of small objects or difficulty manipulating small items like buttons or screws.





Treatment and Management

So far, there is no cure for frontotemporal disorders and no way to slow down or prevent them. 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.

Managing Behavior

The behaviors of a person with bvFTD can upset and frustrate family members and other caregivers. It is natural to grieve for the “lost person,”

but it is also important to learn how to best live with the person he or she has become. Understanding changes in personality and behavior and knowing how to respond can reduce caregivers’ frustration and help them cope with the challenges of caring for a person with a frontotemporal disorder.

Managing behavioral symptoms

can involve several approaches. To

Changing the schedule

ensure the safety of a person and

Matthew, 53, diagnosed

his or her family, caregivers may

with bvFTD, insisted

on playing the card

have to take on new responsibili-

game solitaire on the

ties or arrange care that was not

computer for hours

needed before. For example, they

every morning . He did

not care that this activity interfered with

may have to drive the person to

his wife’s schedule . His wife figured out

appointments and errands, care

how to rearrange her day to stay home

for young children, or arrange for

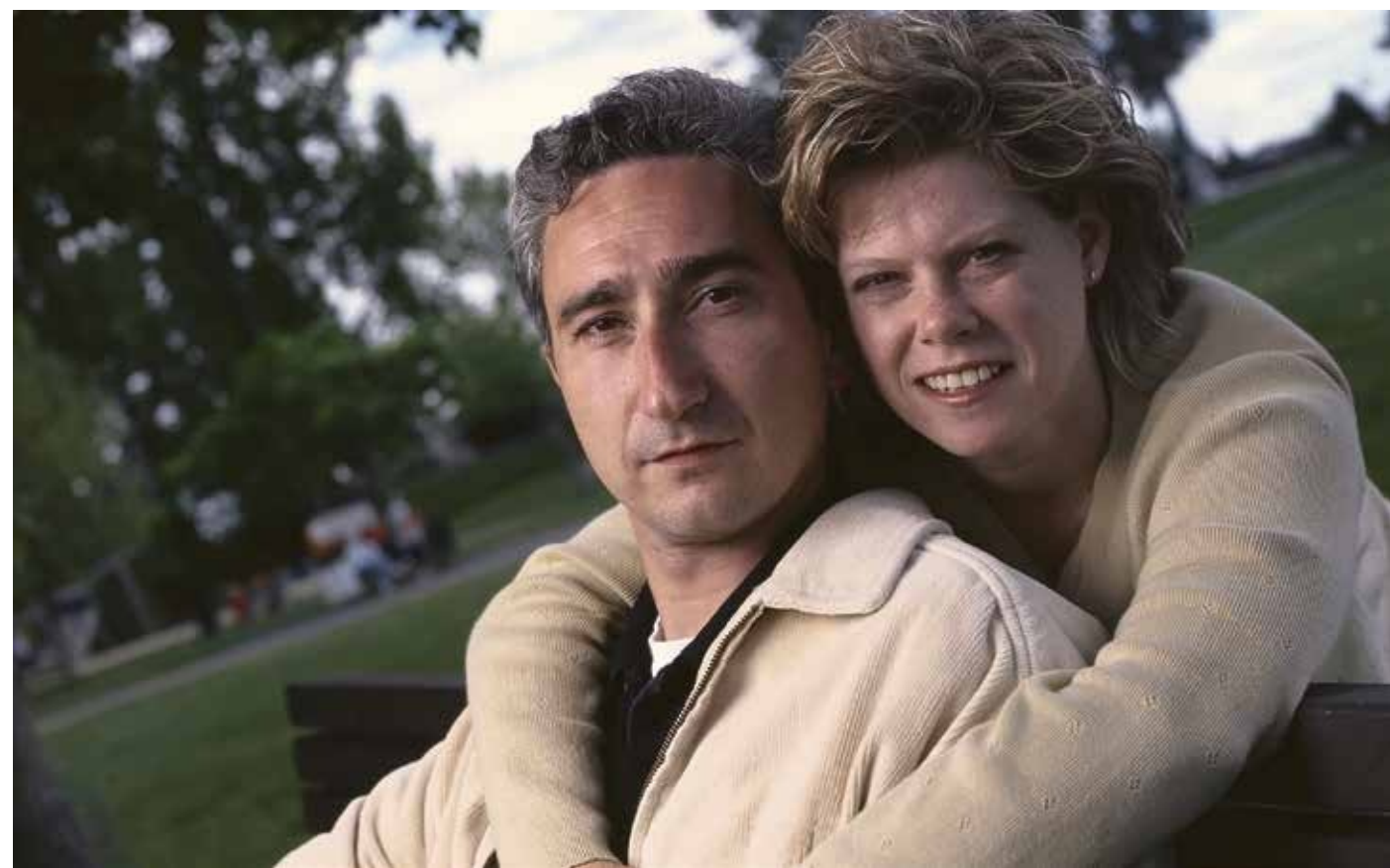
in the morning and take Matthew on

help at home.

errands and appointments in the afternoon . A portable device for solitaire in the car helped distract him .

It is helpful, though often difficult, to accept rather than challenge people with behavioral symptoms.

Arguing with them or talking logically will not help because they cannot control their behaviors or even see that they are unusual or upsetting to others. Instead, be as sensitive as possible and understand that it's the 18



illness “talking.” Frustrated caregivers can take a “timeout”—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 (“What would you like to do today?”) are more difficult to answer than specific ones (“Do you want to go to the movies or the shopping center today?”).

Maintaining the person's schedule and modifying the environment can also help. A regular schedule is less confusing and can help people sleep better.

If compulsive eating is an issue, caregivers may have to supervise eating, limit food choices, lock food cabinets and the refrigerator, and distract the person with other activities. To deal with other compulsive behaviors, caregivers may have to change schedules or offer new activities.

Medications are available to treat certain behavioral symptoms. Antidepressants called selective serotonin reuptake inhibitors are commonly prescribed to treat social disinhibition and impulsive behavior. Patients with aggression or delusions sometimes take low doses of antipsychotic medications. The use of Alzheimer's disease medications to improve behavioral and cognitive symptoms in people with bvFTD and related disorders is being studied, though results so far have been mixed, with some medications making symptoms worse.







Treating Language Problems

Treatment of primary progressive aphasia (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.

To communicate without talking, a person with PPA may use a communication notebook (an album of photos labeled with names of people and objects), gestures, and drawings. Some people find it helpful to use or point to lists of words or phrases stored in a computer or personal digital assistant.

Caregivers can also learn new

Finding a new way to

ways of talking to someone with

communicate

PPA. For example, they can speak

Mary Ann, a television

slowly and clearly, use simple

news anchor for 20

sentences, wait for responses, and

years, began having

trouble reading the

ask for clarification if they don’t

nightly news . At first,

understand something.

her doctor thought

she had a vision problem, but tests showed that her eyesight was normal .

A speech-language pathologist who knows about PPA can test a person's language skills and determine the best tools and strategies to use. Note that many speech-language pathologists are trained to treat aphasia required a medical exam . Her symptoms caused by stroke, which requires different strategies from those used diagnosed logopenic PPA . A speech therapist taught Mary Ann to use a personal digital assistant to express words and phrases . For emergencies, speech-language pathologists and

Mary Ann carries a card in her wallet

that explains her condition .

other experts who know about

frontotemporal disorders.)

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Managing Movement Problems

No treatment can slow down or stop frontotemporal-related movement disorders, though medications and physical and occupational therapy may provide modest relief.

For people with corticobasal syndrome (CBS), movement difficulties are sometimes treated with medications for depression, Alzheimer’s disease, or Parkinson’s disease. But these medicines do not often lead to much improvement. Physical and occupational therapy can help a person with CBS move more easily. Speech therapy may delay the worsening of language symptoms.

For people with progressive supranuclear palsy (PSP), sometimes Parkinson’s disease drugs provide temporary relief for slowness, stiffness, and balance problems. Although physical therapy generally does not help, 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 trouble looking up or down, bifocals or special glasses called prisms are sometimes prescribed.

People with FTD-ALS typically decline quickly over the course of 2 to 3 years. During this time, physical therapy can help treat muscle symptoms, and a walker or wheelchair is 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.

For any movement disorder caused by FTLT, a team of experts can help patients and their families address difficult medical and caregiving issues.

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

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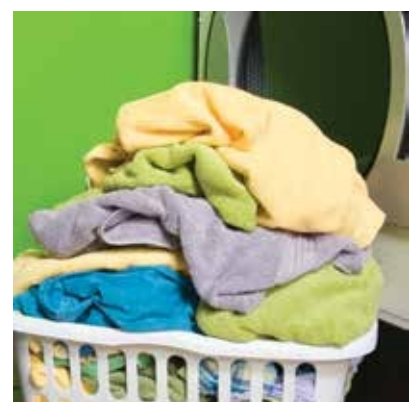
The Future of Treatment

Researchers are looking for better treatments for frontotemporal disorders.

Possible therapies that target the abnormal proteins found in the brain are being tested in the laboratory and in animals. Clinical trials and studies in humans are testing a number of possible treatments. Scientists hope that these studies will lead to the development of new medicines.

Participating in clinical trials is an effective way to help in the fight against frontotemporal disorders. People with frontotemporal disorders and healthy people may be able to take part. To find out more about clinical trials, talk to your health care provider or visit www.ClinicalTrials.gov.





Caring for a Person with a
Frontotemporal Disorder

In addition to managing the medical and day-to-day care of people with frontotemporal disorders, caregivers can face a host of other challenges.

These challenges may include changing family relationships, loss of work, poor health, decisions about long-term care, and end-of-life concerns.

Family Issues

People with frontotemporal disorders and their families often must

Changing relationships

cope with changing relationships,

After Justin gradu-

especially as symptoms get worse.

ated from college,

For example, the wife of a man

he went home to live

with bvFTD not only becomes

with his parents for a

short time . It didn't

her husband's caregiver, but takes

take long to notice

on household responsibilities he

personality changes in his 50-year-old

can no longer perform. Children

mother, a successful executive . She

became more childlike and had trouble

may suffer the gradual "loss" of a

finishing household chores . By the

parent at a critical time in their
time she was diagnosed with bvFTD,
lives. The symptoms of bvFTD
Justin's relationship with his mother
had deteriorated . Learning about the
often embarrass family members
disorder helped Justin understand and
and alienate friends. Life at home
accept the changes he was seeing in
can become very stressful.
his mother .

Work Issues

Frontotemporal disorders disrupt basic work skills, such as organizing, planning, and following through on tasks. Activities that were easy before the illness began might take much longer or become impossible. People lose their jobs because they can no longer perform them. As a result, the caregiver might need to take a second job to make ends meet—or reduce hours or even quit work to provide care and run the household.





Workers diagnosed with bvFTD, Pick's disease, PPA, or ALS can qualify quickly for Social Security

disability benefits through the “compassionate allowance” program. For more information, see www.socialsecurity.gov/

compassionateallowances or call 1-800-772-1213.

Caregiver Health and Support

Caring for someone with a frontotemporal disorder can be very hard, both physically and emotionally. To stay healthy, caregivers can do the following:

- Get regular health care.
- Ask family and friends for help with child care, errands, and other tasks.
- Spend time doing enjoyable activities, away from the demands of caregiving. Arrange for respite care—short-term caregiving services that give the regular caregiver a break—or take the person to an adult day care center, a safe, supervised

environment for adults with

A stressful disorder

dementia or other disabilities.

Robert, 60, started

- Join a support group for

humming and

whistling through-

caregivers of people with

out the day . Over

frontotemporal disorders.

time, his humming

Such groups allow caregivers

got louder, and he

started banging on the table over and

to learn coping strategies and

over again . Robert’s wife and his boss

share feelings with others in
at work were concerned about this
the same position.
behavior and tried to distract him, but
this became stressful for everyone .
The organizations listed in the
Robert lost his job . A doctor diagnosed
bvFTD and prescribed medication to
Resources section of this booklet
control his behavior . Robert began to
(starting on page 27) can help
attend an adult day services program,
with information about caregiver
but his behavior was too hard for the
staff to manage, and he had to leave
services and support.
the program .



Long-Term Care

For many caregivers, there comes a point when they can no longer take care of the person with a frontotemporal disorder without help. The caregiving demands are simply too great, perhaps requiring around-the-clock care. As the disease progresses, caregivers may want to get home health care services or look for a residential care facility, such as a group home, assisted living facility, or nursing home. The decision to move the person with a frontotemporal disorder to a care facility can be difficult, but it can also give caregivers peace of mind to know that the person is safe and getting good care.

End-of-Life Concerns

People with frontotemporal disorders typically live 6 to 8 years with their conditions, sometimes longer, sometimes less. Most people die of problems related to advanced disease. For example, as movement skills decline, a person can have trouble swallowing, leading to aspiration pneumonia, in which food or fluid gets into the lungs and causes infection. People with balance problems may fall and seriously injure themselves.

It is difficult, but important, to plan for the end of life. Legal documents, such as a will and durable powers of attorney for health care and finances, should be created or updated as soon as possible after a diagnosis of bvFTD, PPA, or a related disorder. Early on, many people can understand and participate in legal decisions. But as their illness progresses, it becomes harder to make such decisions.

A physician who knows about frontotemporal disorders can help determine the person's mental capacity. An attorney who specializes in elder law, disabilities, or estate planning can provide legal advice, prepare documents, and make financial arrangements for the caregiving spouse or partner and dependent children. If necessary, the person's access to finances can be reduced or eliminated.

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Conclusion

It is impossible to predict the exact course of frontotemporal disorders.

These disorders are not easy to live with, but with help, people can meet the challenges and prepare for the future. Getting an early, accurate diagnosis and the right medical team are crucial first steps. Researchers and clinicians are working toward a deeper understanding of frontotemporal disorders and better diagnosis and treatment, which may help people manage these difficult conditions.

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Resources

Federal Government

National Institute on Aging

Alzheimer's Disease Education and Referral (ADEAR) Center P.O. Box 8250

Silver Spring, MD 20907-8250

1-800-438-4380 (toll-free)

www.nia.nih.gov/alzheimers

The National Institute on Aging (NIA), part of the National Institutes of Health, leads the Federal Government effort conducting and supporting research on Alzheimer's disease and other forms of dementia. NIA's Alzheimer's Disease Education and Referral (ADEAR) Center offers information and publications on dementia and caregiving for families, caregivers, and professionals. Free publications include: *Getting Your Affairs in Order*, *Nursing Homes: Making the Right Choice*, *End of Life: Helping with Comfort and Care*, and many others.

National Institute of Neurological Disorders and Stroke NIH Neurological Institute

P.O. Box 5801

Bethesda, MD 20894

1-800-352-9424 (toll-free)

1-301-468-5981 (TTY)

www.ninds.nih.gov

MedlinePlus

National Library of Medicine

www.medlineplus.gov

Organizations

Association for Frontotemporal Degeneration

Radnor Station Building 2, Suite 320

290 King of Prussia Road

Radnor, PA 19087

1-267-514-7221

1-866-507-7222 (toll-free)

www.theaftd.org

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ALS Association

27001 Agoura Road, Suite 250

Calabasas Hills, CA 91301-5104

1-800-782-4747 (toll-free)

www.alsa.org

CurePSP

30 East Padonia Road, Suite 201

Timonium, MD 21093

1-800-457-4777

www.curepsp.org

National Aphasia Association

350 Seventh Avenue, Suite 902

New York, NY 10001

1-800-922-4622 (toll-free)

www.aphasia.org

WE MOVE

5731 Mosholu Avenue

Bronx, NY 10471

www.wemove.org

Caregiver Services

Eldercare Locator

1-800-677-1116 (toll-free)

www.eldercare.gov

Family Caregiver Alliance

180 Montgomery Street, Suite 900

San Francisco, CA 94104

1-800-445-8106 (toll-free)

www.caregiver.org

National Academy of Elder Law Attorneys, Inc.

1577 Spring Hill Road, Suite 220

Vienna, VA 22182

1-703-942-5711

www.naela.org

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Caregiver Support

FTD Support Forum

www.ftdsupportforum.com

PPA Support Group

www.groups.yahoo.com/group/PPA-support

Well Spouse Association

63 West Main Street, Suite H

Freehold, NJ 07728

1-800-838-0879

www.wellspouse.org

Diagnosis, Treatment, and Research Centers

Columbia-Presbyterian Medical Center

Lucy G. Moses Center for Memory and Behavioral Disorders New York, NY

1-212-305-6939

http://pathology.columbia.edu/previous/taub/centers.html Indiana University School of Medicine

Indiana Alzheimer's Disease Center

Indianapolis, IN

1-317-278-5500

<http://iadc.iupui.edu>

Johns Hopkins University School of Medicine

Frontotemporal Dementia and Young-Onset Dementias Clinic Baltimore, MD

1-410-502-2981

www.hopkinsmedicine.org/Psychiatry/specialty_areas/neuropsychiatry/

[frontotemporal_dementia](#)

Massachusetts General Hospital

Frontotemporal Disorders Unit

Boston, MA

1-617-726-5571

www.ftd-boston.org

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Mayo Clinic

Department of Neurology

www.mayoclinic.org/frontotemporal-dementia

Rochester, MN

1-507-538-3270

www.mayoclinic.org/frontotemporal-dementia/rsttreatment.html Jacksonville, FL

1-904-953-0853

www.mayoclinic.org/frontotemporal-dementia/jaxtreatment.html Scottsdale, AZ

1-800-446-2279

www.mayoclinic.org/frontotemporal-dementia/scttreatment.html Northwestern University Feinberg School of Medicine Cognitive Neurology and Alzheimer's Disease Center Chicago, IL

1-312-908-9339

www.brain.northwestern.edu

University of California, Los Angeles

Frontotemporal Dementia & Neurobehavior Clinic Los Angeles, CA

1-310-794-2550

www.adc.ucla.edu/Frontotemporal_Dementia_Clinic.asp University of California, San Francisco

Memory and Aging Center

San Francisco, CA

1-415-476-6880

www.memory.ucsf.edu/ftd

University of Pennsylvania Health System

Center for Frontotemporal Dementia

Philadelphia, PA

1-215-662-3606

www.uphs.upenn.edu/ftd

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National Institute on Aging

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Executive Director

Association for Frontotemporal Degeneration

For additional copies of this publication or further information: www.nia.nih.gov/alzheimers

1-800-438-4380

National Institute on Aging

Alzheimer's Disease Education and Referral Center



U.S. Department of Health and Human Services

National Institutes of Health

National Institute on Aging

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