



Frontotemporal Degeneration: Misdiagnosed and Misunderstood

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No Disclosures

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FTD Overview and Specific FTD Disorders

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Dementia estimates and hallmarks

Alzheimer's Disease

- 5.8 million people in the U.S.
- Memory loss is the most common early symptom
- Amyloid plaques and tangles

Vascular Dementia

- ~10-15% of all dementias are vascular only
- ~40% of all dementias have a vascular component
- Caused by stroke(s) or other cerebrovascular damage

Lewy Body Dementia

- 1.4 million people in the U.S.
- Typically involves motor symptoms in addition to cognitive and behavioral changes
- Alpha-synuclein protein deposits on nerve cells

Frontotemporal Degeneration

- ~60,000 people in the U.S. but commonly misdiagnosed
- Most common forms involve changes in language or behavior
- Younger age of onset
- TAU, TDP-43 and FUS

Mixed dementia ~50%

Alzheimer's Association. 2019; Aarsland et al. 2008; Knopman, D. 2019; Schneider, J. et al. 2007

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FTD: A Cluster of Complex Disorders

also called: frontotemporal dementia, frontotemporal lobar degeneration or Pick's Disease

Progressive Behavior/ Personality Change

Behavioral Variant FTD (bvFTD)

Progressive Language Change

Primary Progressive Aphasia (PPA)

Subtypes:

- Nonfluent/Agrammatic Variant
- Semantic Variant
- Logopenic Variant

Progressive Motor Function/ Movement Change

Corticobasal Syndrome (CBS/CBD)

Progressive Supranuclear Palsy (PSP)

FTD + Motor Neuron Disease (ALS-FTD)

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FTD and the Brain

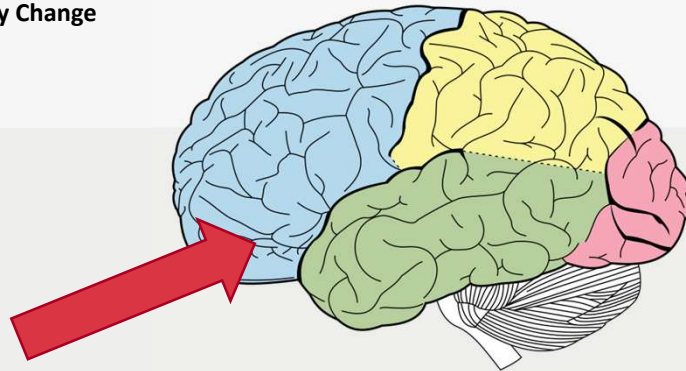
Progressive Behavior/Personality Change

Behavioral Variant FTD (bvFTD)

Frontotemporal dementia

Most common form of FTD!

- Reasoning, decision making
- Control of behavior
- Problem solving
- Attention, concentration
- Emotional control
- Safety awareness
- Initiating action
- Physical movement
- Executive functions (Planning, organizing)



FRONTAL LOBE

Prefrontal cortex

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FTD and the Brain

Progressive Language Change

Primary Progressive Aphasia (PPA)

Subtypes:

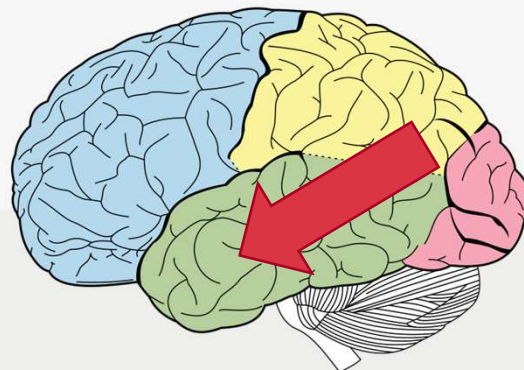
- Nonfluent/Agrammatic Variant
- Semantic Variant
- Logopenic Variant

Right Lobe

- Inhibition of speech
- Visual memory, pictures, shapes and faces, art

Left Lobe

- Verbal memory
- Understanding words and names
- Sorting new information



TEMPORAL LOBE

Processing sensory information

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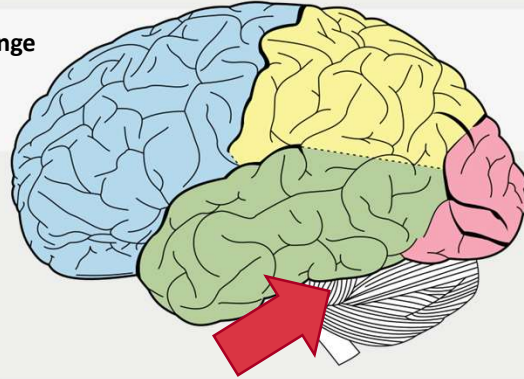
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FTD and the Brain

Progressive Motor Function/ Movement Change

Corticobasal Syndrome (CBS/CBD)
Progressive Supranuclear Palsy (PSP)

- Voluntary movement
- Cognition
- Emotional processing



Basal Ganglia &
Brain Stem

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FTD-ALS

The discovery in 2011 that the *C9orf72* gene mutation can cause both FTD and amyotrophic lateral sclerosis (ALS) has transformed a long-held belief that ALS is 'purely' a movement disorder and that FTD is 'purely' a cognitive or behavioral form of dementia.



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Is FTD Genetic?

SPORADIC FTD

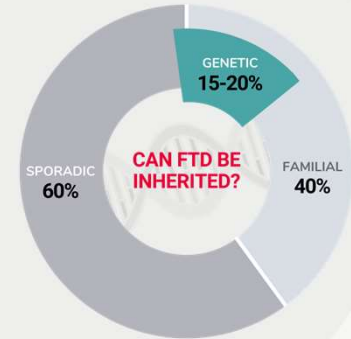
- For most people with FTD no one else in the family has FTD. The *presumption* is that relatives of the person diagnosed do not have an increased risk.
- * *Approximately 5-10% of individuals with no family history of FTD have an identifiable genetic cause; a new “de novo” change.*

FAMILIAL FTD

- Approximately 40% of people with FTD have a family history of dementia, a major psychiatric condition, or progressive changes in movement.

GENETIC FTD

- A subset of familial FTD cases are caused by a variant in a single gene inherited from a parent.



AFTD strongly recommends genetic counseling as a first step to answer questions about genetic status.

For more info: www.theaftd.org/ftd-genetics/what-causes-ftd/

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Impact on Families

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Economic & Social Costs

\$99k
 ↓
\$50k

Twelve months before an FTD diagnosis, most families reported a household income in the \$75,000–\$99,000 range. Twelve months after diagnosis, income fell as much as 50 percent.

FTD vs **ALZ**

Overall, families dealing with FTD face an economic burden of around

\$120,000

each year — roughly twice the economic burden of Alzheimer’s.

37%

of FTD caregivers said they stopped working post-diagnosis.

58%

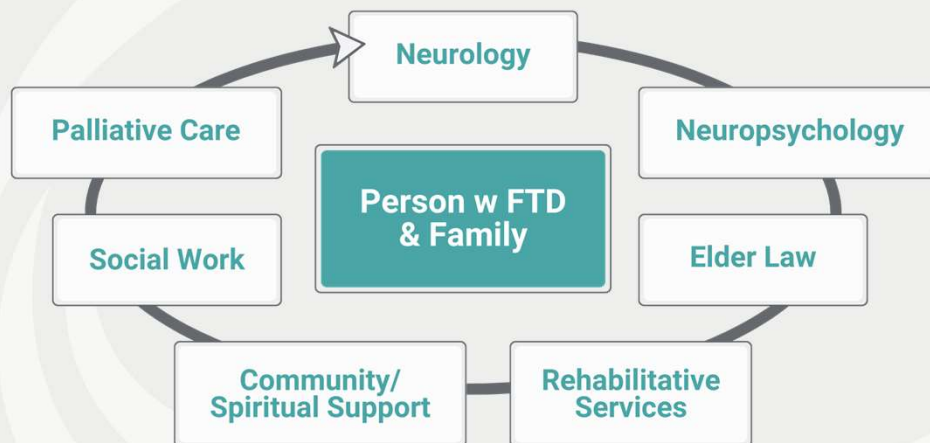
of respondents said that FTD caused their loved ones to make poor financial decisions.

**Published in the scientific journal Neurology, 11/14/17*

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Helping Families Build a Care Team



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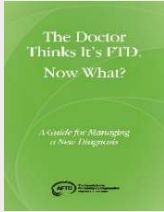
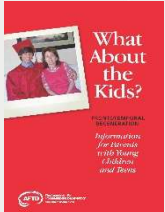
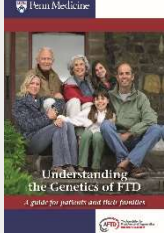
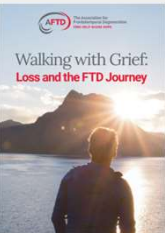
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Resources from AFTD

Helpline	info@theaftd.org 866-507-7222
Websites	www.theaftd.org www.aftdkidsandteens.org
Publications	<i>Help & Hope</i> <i>Partners in FTD Care</i> <i>The Doctor Thinks It's FTD. Now What?</i> <i>What About the Kids?</i> <i>Understanding the Genetics of FTD</i> <i>Walking with Grief: Loss and the FTD journey</i>
Grants	Respite, Travel, Quality of Life
Support	National and regional on-line and local in-person groups theaftd.org/living-with-ftd/aftd-support-groups

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AFTD Resources for Medical and Care Professionals

- [Diagnostic Checklists](#): for bvFTD and PPA
- Partners in Care quarterly publication focusing on a specific issue or topic
 - [Partners in FTD Care | AFTD \(theaftd.org\)](#)
- Clinical presentations of symptoms:
 - [Clinical Presentations | AFTD \(theaftd.org\)](#)
- Webinars and Annual Conference
- AFTD Helpline: 1-866-507-7222 or info@theaftd.org

For additional info, go to [For Health Professionals | AFTD \(theaftd.org\)](#) or contact the AFTD Helpline.

