



THE MANY MANIFESTATIONS OF FRONTOTEMPORAL DEMENTIA (FTD)

CHRISTINE J. CLIATT BROWN, MD
ASSISTANT PROFESSOR | COGNITIVE NEUROLOGY
UNIV. OF UTAH NEUROLOGY & GEORGE E. WAHLEN VETERAN'S AFFAIRS

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OVERVIEW

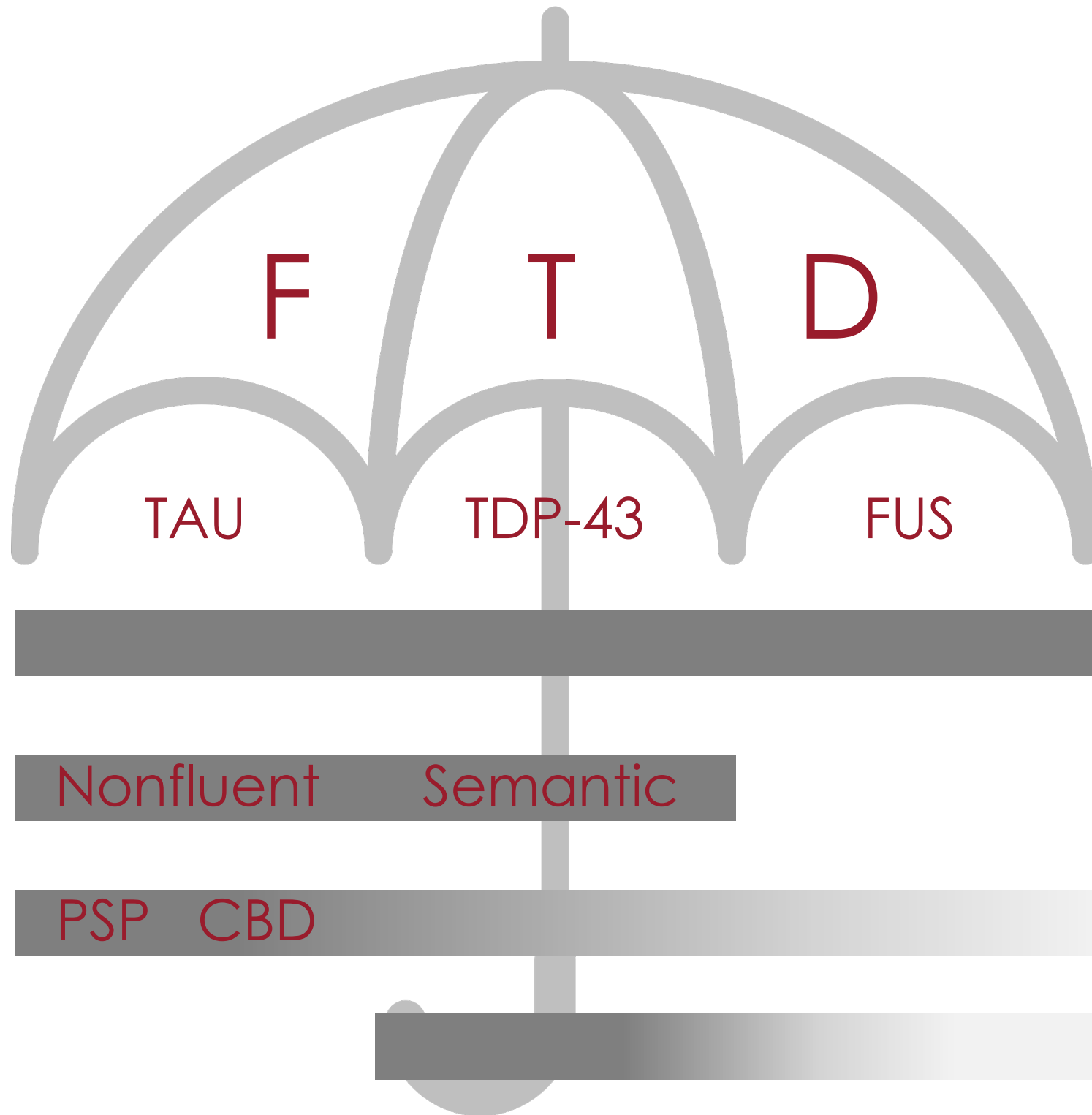
- What is dementia?
- FTD Umbrella
- Behavioral Variant FTD
- Primary Progressive Aphasia
- FTD Parkinson's Plus
- FTD-ALS
- Diagnosis
- Management

WHAT IS DEMENTIA?

- **Dementia:** cognitive decline impacting ability to function fully independently
- **Mild Cognitive Impairment (MCI):** cognitive decline with preserved independent functioning
- Descriptive terms that do not describe the underlying cause
- Degenerative diseases: Alzheimer's Disease, FTD, DLB, and others
- Cognition:
 - Memory
 - Speech, language
 - Executive function
 - Reasoning, judgment
 - Visual function
 - Social & behavioral
 - Attention, processing speed

FTD UMBRELLA

- Degenerative diseases that affect the frontal and/or temporal lobes
- Younger age of onset, before age 60
- FTD is more often familial than AD
 - Up to 40% familial (especially bvFTD)
 - 12-30% with identifiable genetic causes
- Varying pathologies:
 - Tau
 - TDP-43
 - FUS
 - Others
- Varying symptoms or clinical syndromes
 - bvFTD
 - PPA
 - Parkinson's Plus Syndromes: PSP, CBD
 - FTD-ALS
 - Other



CASE 1 BEHAVIORAL VARIANT FTD

58-year-old man with progressive changes in personality and behavior

- Making rude comments to family, even strangers
- Impulsive purchases – bought a boat without consulting his wife (they don't live near water)
- Binge-eating salt & vinegar potato chips and uncooked hot dogs
- Loss of motivation – not doing chores, doesn't want to socialize and has abandoned his hobbies
- Sits watching the same western movies over and over
- Worsening steadily, recently unbuttoned his pants and licked his plate at a nice restaurant
- Having trouble using phone and computer

FTD BEHAVIORAL VARIANT

- Progressive decline in behavior, social comportment, and cognition with changes in behavior being EARLY features
- Cognition: executive function deficits with relative sparing of memory and visuospatial function
- Behavioral disinhibition
- Apathy or inertia
- Loss of sympathy or empathy
- Perseverative, compulsive behaviors
- Hyperorality, dietary changes

CASE 2 PRIMARY PROGRESSIVE APHASIA

64-year-old woman with progressive changes in language

- Word-finding difficulty
- Spelling errors – spelled scissors “sissers”
- Difficulty recalling names, even of very familiar people
- Having a hard time communicating her thoughts, often using generic words like “thing,” which makes it hard for others to understand her
- Son has to make phone calls for her
- Recently, not understanding words she should know (pomegranate, cardigan)
- On language testing, she could not name dinosaur and thought it would be found in a zoo

FTD PPA

Primary Progressive Aphasia: A dementia of the language network. Speech and language problems are the first and most prominent deficits.

SEMANTIC

- Word-finding difficulty
- Difficulty understanding the meaning of words
- Loss of knowledge about words, objects, animals, etc.
- Changes in spelling

NONFLUENT

- Loss of normal grammar
- Effortful, halting speech with mispronunciation
- Difficulty understanding more complex or oddly worded sentences

CASE 3 PROGRESSIVE SUPRANUCLEAR PALSY

74-year-old man with frequent falls, difficulty walking, and progressive behavior changes

- Falls began three years ago, now falling 2-4 times per week
- Often falls back when trying to rise from a chair
- Slow, stiff, and shuffling when walking
- Sometimes freezes up when walking
- Says he can't see his feet when going down the stairs
- Seems to have "lost his filter"
- Can not figure out how to fix things around the house
- No longer able to manage the finances, has been spending excessively
- Voice is soft and speech is slurred, occasional choking on food

FTD PARKINSON'S PLUS

- Parkinsonism – motor changes including slow movement, rigidity, tremor, shuffling gait
- Progressive Supranuclear Palsy (PSP): early frequent falls, eye movement changes, slurred speech, cognitive & behavioral decline
- Corticobasal Degeneration: asymmetric apraxia, rigidity, other features
- Apraxia: inability to perform learned movements, tasks, or gestures
- Parkinsonism can also be a feature in non-PSP/CBD FTD

⁴Höglinger GU, Respondek G, Stamelou M, et al. Clinical diagnosis of progressive supranuclear palsy: The movement disorder society criteria. *Mov Disord.* 2017;

⁵Armstrong MJ, Litvan I, Lang AE, et al. Criteria for the diagnosis of corticobasal degeneration. *Neurology.* 2013

FTD ALS

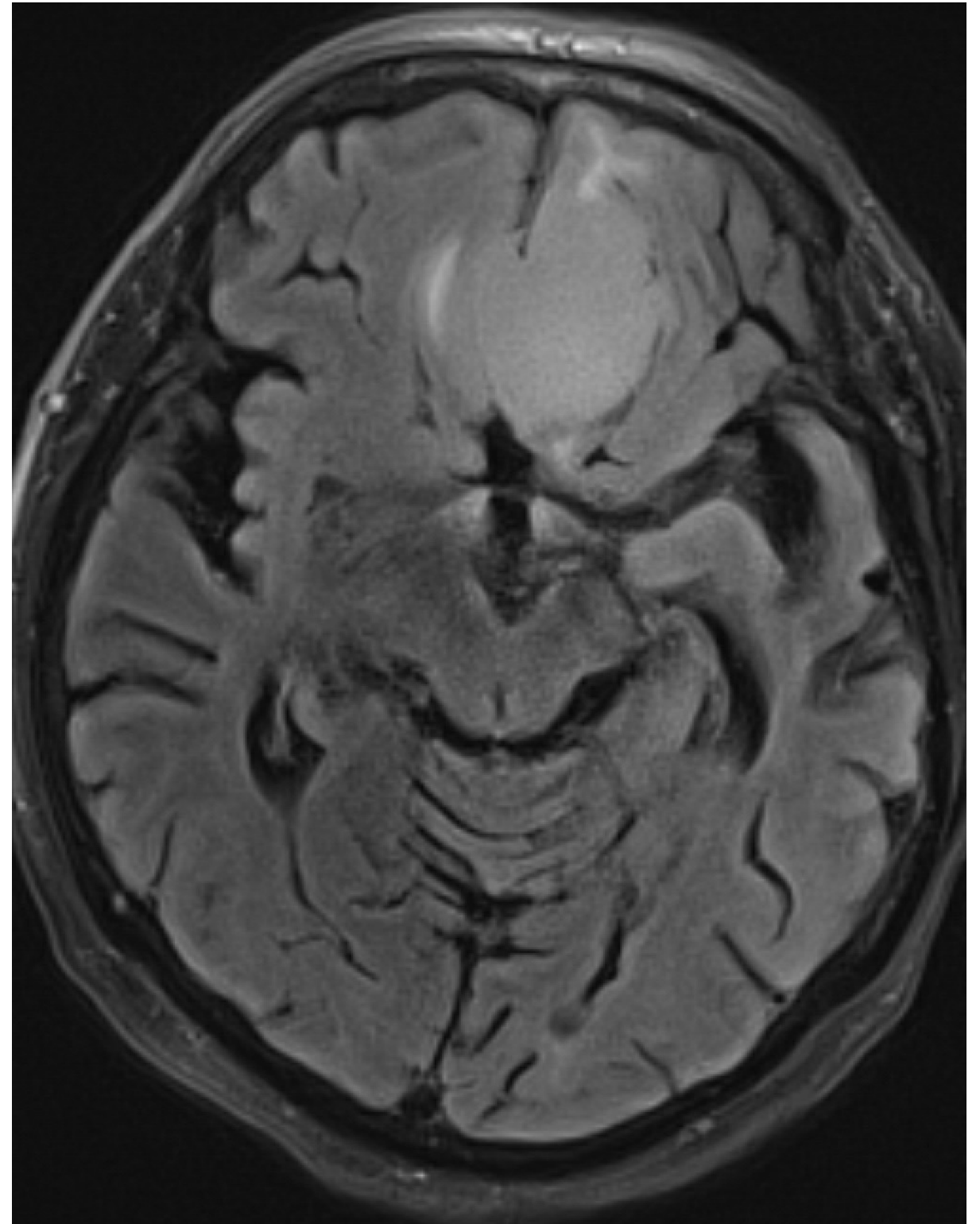
- Amyotrophic Lateral Sclerosis (Lou Gherig Disease) – progressive brain and spinal cord disease
- Weakness
- Difficulty walking, wheelchair bound
- Difficulty speaking and swallowing
- Life expectancy 2-5 years
- Around 15% with FTD develop ALS
- Around 20% with ALS develop FTD, 50% developing cognitive or behavioral symptoms
- C9orf72 repeat expansion increases risk of FTD and ALS
- ALS diagnosed with exam and EMG

⁶Cividini C, Basaia S, Spinelli EG, et al. Amyotrophic Lateral Sclerosis-Frontotemporal Dementia: Shared and Divergent Neural Correlates Across the Clinical Spectrum. *Neurology*. 2022

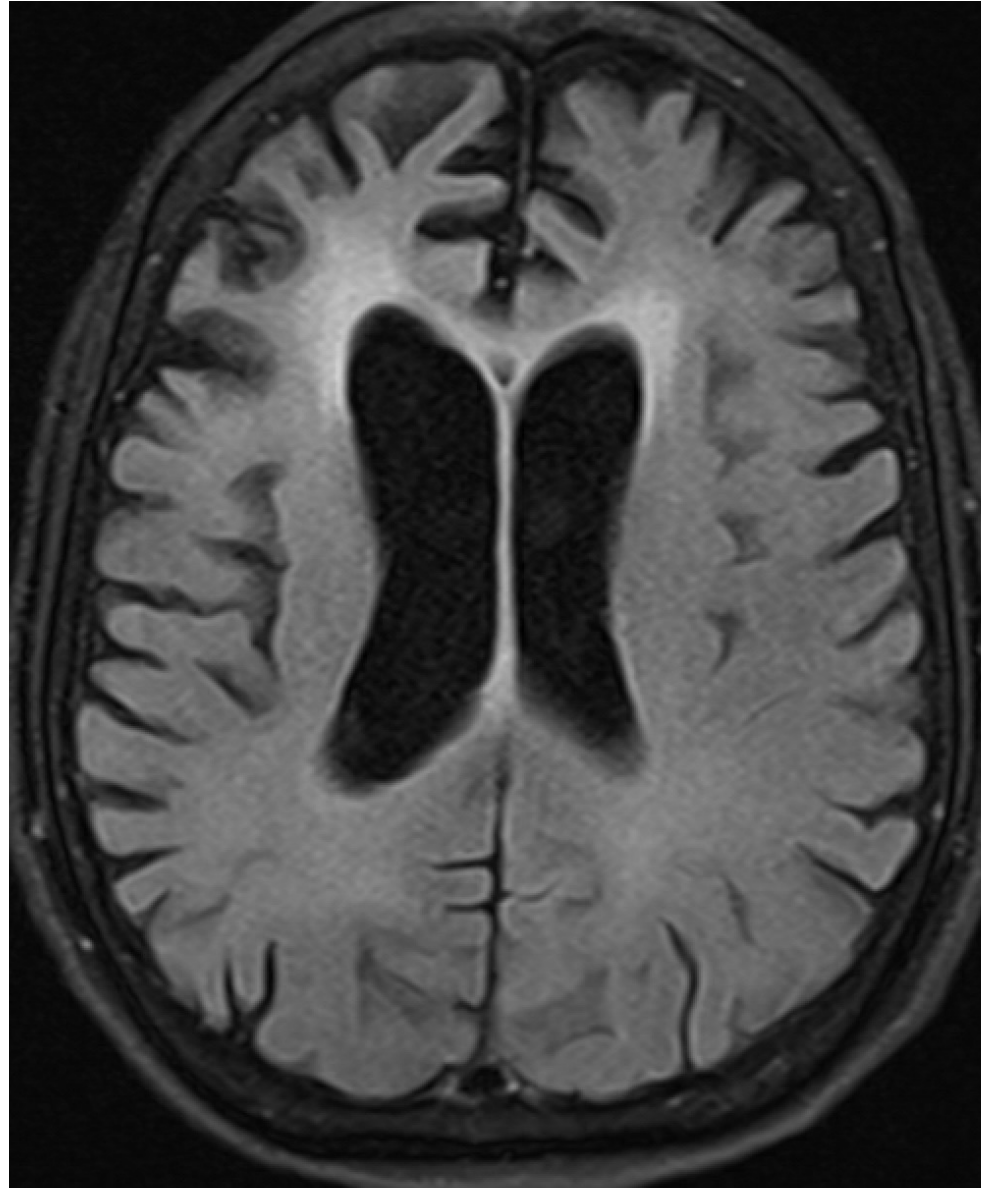
FTD DIAGNOSIS

Why get testing?

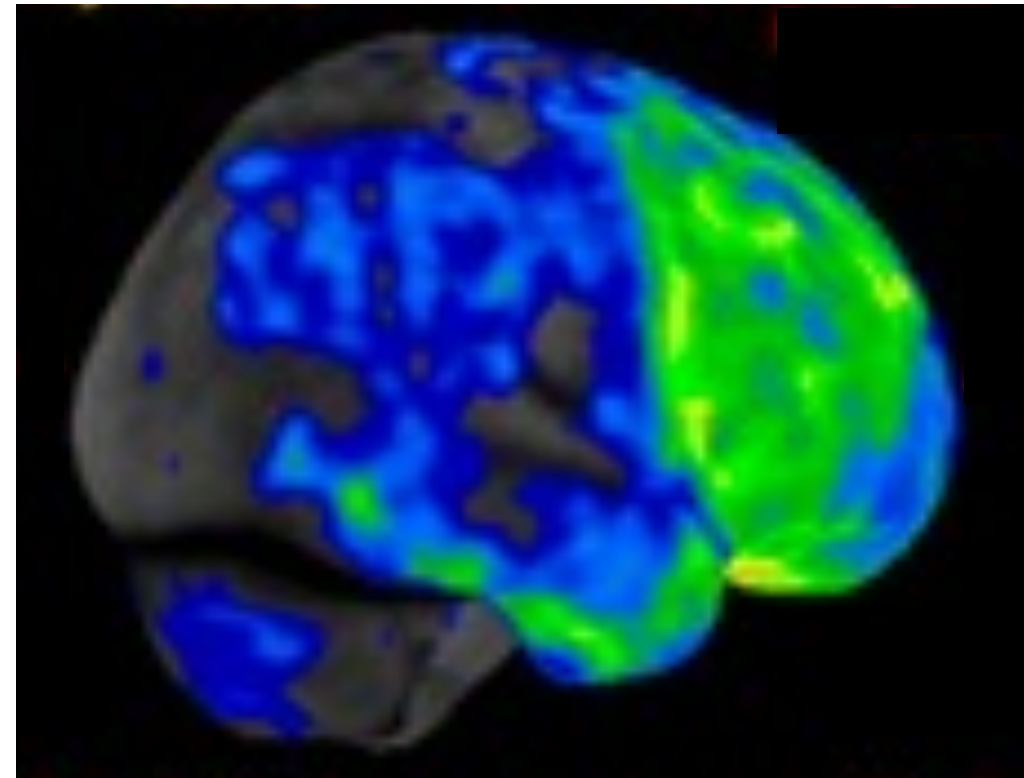
- Understanding
- Excluding treatable causes (tumor, seizures, autoimmune causes)
- Medication plans
- Understand genetic risk
- Recognizing ALS risk
- Clinical trials, studies
- Prognosis
- Symptom progression
- Appropriate therapies



FTD DIAGNOSIS



- Neuropsychological testing
- MRI
- FDG PET
- Ruling out Alzheimer's Disease
 - Spinal fluid biomarkers
 - Amyloid PET
- Genetic testing



FTD MANAGEMENT

- No medications can stop or slow progression in FTD
- Ongoing research
- Some medications can help manage behavioral symptoms
- Physical therapy and occupational therapy for those with motor symptoms
- Speech therapy for those with PPA
- Understanding resources
- Nonpharmacologic management
 - Caregiver education
 - Communication strategies
 - Lifestyle

MANAGING BEHAVIOR

- Routine
 - Sleep schedule
 - Meals
 - Exercise
 - Activities/hobbies
- Recognize and avoid triggers
- Give simple choices (bath or shower)
- Use a calm, positive tone; smile, laugh
- Validate feelings, don't try to reason
- Don't point out deficits
- Provide distractions
- Use rewards
- Look for (and treat) reasons for worsening
 - Pain
 - Constipation
 - Insomnia
 - Depression, anxiety
 - Infection
 - Medication side effect
- Take breaks
- Know when to get help

HOW TO SEEK EVALUATION

- Start with your PCP
 - Cognitive screening test
 - Blood work
 - MRI
 - Look for treatable causes (sleep apnea, insomnia, depression, medication side effect, etc.)
- Referral to a specialist:

Cognitive Disorders Clinic

175 N Medical Dr

Salt Lake City, UT 84132

Request an appointment at **801-585-7575**

FTD RESOURCES

[Alzfdn.org/caregiving-resources](https://alzfdn.org/caregiving-resources)

theAFTD.org

FTDregistry.org

PSP.org

Caregiver.org

Aphasia.org



Find Your Local Aging Services

In-home services, and support and classes for caregivers



Alz Assoc. Resource Finder, including support groups

800-272-3900



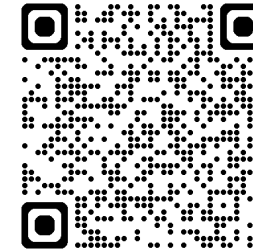
Alz.connect

Caregiver Forum. A place to find answers from other caregivers



Seniors Blue Book

Directory of Services for Seniors



Alzheimer's Foundation of America

Resources for Alzheimer's and Dementia Caregivers



Book, "Creating Joy Along the Alzheimer's Journey"

(helpful for all types of dementia care)

<https://www.amazon.com/Creating-Moments-Along-Alzheimers-Journey/dp/1557537607>



Alzheimer's Association (Supporting all types of dementia)

<https://training.alz.org/>



National Institute on Aging

[Order a Free Manual: Caring for a Person with Alzheimer's.nia.nih.gov/publication](https://www.nia.nih.gov/publication)



Dementia Care with Teepa Snow

Podcasts: [dementia-care-partner-talk-show](https://www.dementia-care-partner-talk-show.com/)

Questions?



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