



Introduction to FTD: Advancing Help and Hope

Kansas Alzheimer's Association Education Conference
Sharon S. Denny, Sr. Director of Programs, AFTD | Nov. 14, 2019

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Objectives

- Identify the main FTD syndromes and the importance of accurate diagnosis.
- Identify ways that caregiving for people with FTD may differ from caring for someone with Alzheimer's disease.
- Learn about specialized resources for support, education and advocacy to improve care for people facing FTD.

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Frontotemporal Degeneration (FTD)

Progressive disease of frontal and/or temporal lobes

Most common dementia under age 60

- Age of onset: 20-80 (av. 57)

Prevalence: 50,000-60,000 in the US
~20 per 100,000 (45-64 y.o)

Various clinical syndromes:

- Behavior, language and movement changes
- Not primarily memory disorder

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Heterogeneity

FTD is a complex group of overlapping disorders

- Wide range of symptoms and expressions of them
- Multiple pathologies (tau, TDP-43, FUS)
- Complex genetics (*MAPT*, *GRN*, *C9orf72*)
- Variable rates of progression

Each person's experience of FTD is as individual as they are.

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Neuropathology

"Frontotemporal lobar degeneration" (FTLD) - the specific pathological diseases that result in FTD syndromes"

Subtypes are based on the proteins found within neuronal inclusions. Most are either:

- FTLD-tau (Pick's disease, some PPA, CBD and PSP)
- FTLD-TDP (some bvFTD, svPPA, ALS/FTD)

In addition to FTLD pathology PPA can be caused by AD:



FTLD-T



TDP - 43



Alzheimer's Pathology

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Images courtesy N. Cummings

Genetics in FTD

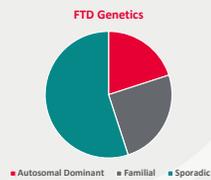
Genetic causes play greater role than in Alzheimer's

Etiology

- ~ 15-20% dominantly inherited
- ~ 20-30% familial
- ~ Half are sporadic

3 main genes:

MAPT, *GRN*, *C9orf72*
Other rare mutations



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FTD vs Alzheimer's Disease

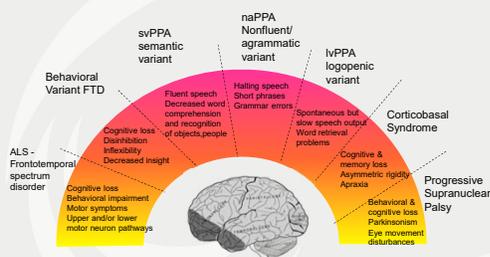
	FTD	Alzheimer's
Age of onset	Av. 50's-60's	> 65; av. ~ 80
Prevalence in US	50,000-60,000	5.8 million
Clinical hallmarks	Behavior, language, movement	Memory loss
% Inherited	10-20%	< 1%
Time to diagnosis	3.6 years	2.8 years

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FTD Syndromes and Diagnosis

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FTD is a Spectrum of Diseases



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Cognitive Changes

Executive Functions

Attention

- Can't watch movies, read books, converse at length
- Appear purposeless and bored (apathetic)

Making and carrying out plans

- Work difficulties - problems with planning, organization, feedback, correction, task completion
- Trouble paying bills, cooking, shopping, grooming

Reasoning, flexible thinking

- Mental rigidity, insist of having things a certain way

Memory is relatively spared

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Behavioral variant FTD (bvFTD)

Changes in behavior and personality

Behavioral Disinhibition

- Socially inappropriate behaviors
- Loss of manners or decorum
- Impulsive, rash, careless actions

Apathy

Loss of empathy

Perseverative, compulsive behaviors

Changes in diet & oral behaviors

Impaired judgment, foresight, reasoning

A disorder of social comporment

Rascovsky et al Brain on line Aug 2, 2011 © 2014 AHA 11

What you may see:

Changes in "Social Brain"

- Loss of interest in family, friends
- Lack of concern over family illness/death; disinterest in daughter's prom
- Increase in self-centered actions
- Impulsive spending (5 new cars)
- Vulnerable to scams
- Driven to touch or use items in their view- touch a stranger's hair, take food when not hungry
- Delusions - jealousy, religious, unusual thoughts



Image - DiAber123

"Who is that man in this house?"

"...this is not the same person I married"

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ALS-FTD Spectrum Disorder

Most common genetic cause of ALS & FTD

Cognitive impairment present in many with ALS

- Small portion meet criteria for bvFTD or PPA
- *C9orf72* gene chr 9
- Accounts for ~11% familial FTD cases
- Muscle weakness, muscle atrophy of ALS with bvFTD or PPA symptoms
- Often rapid progression
- Especially complex care needs

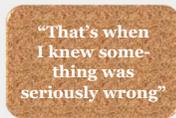


FTD Diagnostic Challenges

Unexplained changes - Long evaluation process

Gradual onset; younger age

- Early symptoms dismissed
- Try to resolve confusion—stress, anxiety, relationship issues?
- Treated as depression, bipolar disorder, Parkinson's, etc
- Manage without help; antagonism
- "Tipping point" prompts further action



FTD Diagnostic Challenges

Clinical diagnosis based on history & examination

Currently no FTD biomarkers

- History - critical to show change from prior functioning
- Neuropsychological testing
- Brain imaging (MRI or PET scan)
- CSF studies are generally not helpful, except to diagnose AD
- Genetic testing should not be used for diagnosis, rather for counseling once a diagnosis has been made

FTD presents distinct care and support needs.

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Impact on Family System

Most common dementia under 60

- Coping before diagnosed
- Relationships strained or broken
- Loss of employment, often peak of career
- Still physically robust and active
- Fewer co-morbid health conditions



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Impact on Family System

Feeling isolated magnifies challenges

- Roles change-
- Young adult and school age children
- Behaviors intrusive, embarrassing
- Friends pull away
- Ambiguous loss



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Economic Burden Study*

\$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

Care Management Challenges

No FTD treatments or clinical care guidelines

- Not known to many physicians & health providers
- Few experienced home & community services
- Eligibility under 65 yrs
- Behavior issues increase care burden
- No medications for cognitive symptoms (eg. awareness, judgment)

The Experience of FTD

Diverse Clinical Symptoms

Younger Onset

Less Common

GREATER CARE NEEDS

Diagnostic Challenges

Family & Financial Impact

Complex pathology & genetics

Isolation and “care burden” are higher

What can we do?

FTD Families Require

Disease Education

Individualized & Coordinated Care

Help & Hope

FTD Peer Support

Awareness of Research

Advocacy

Personal Attention
Explore the family’s experience

Diagnostic experience can shape subsequent contacts

- With doctors, providers, family & researchers
- Frustration, guilt, anger
- Divorce before diagnosis

Post diagnosis sense-making:

- Expand concept of dementia (younger, mobile)
- Reframe pre-diagnostic phase (guilt, hurt)
- Re-frame relationships

(K. Rogers, presentation, ICFTD 2018)

Disease Education

The best intervention is a well-informed & supported person with FTD and care partner

- FTD symptoms, subtypes and value of diagnosis
- Disease process (symptoms beyond person's control)
- Observing & describing behaviors
- Pathology, genetics and potential family risk
- Importance of research and emerging clinical trials



FTD-specific Support

Support from those who understand is critical

- Peer-support (face to face, phone, online)
- Ambiguous loss – physically here; seems like stranger
- Creative problem-solving assistance
- Identification of local resources
- Encourage use of individual, child & family counseling

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Quality of Life

Focus on abilities and strengths

- Maximize inclusion of person with FTD
- Adjust expectations
- Maximize engagement
- Do preferred activities - differently
- Assess needs of the family system
- Attend to needs of each member



"We think of a person with dementia only in the advanced stage: almost entirely lost and bewildered, confused, limited, or incapacitated. That's not me!"
-Rev. Tracey Lind

Specialized Care

Alzheimer's approaches are not sufficient in FTD

Non-pharmacological interventions most effective

- Partner with family – need true team approach
- Productive daily routine; speech-language therapy
- Simplified, structured environment
- Intact memory

Assess for safety regularly

Medication for targeted symptoms

- Alzheimer's meds not indicated



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Advocate for Services

Finding appropriate services is difficult

Build a care team

- FTD specialist, physician, palliative care consult
- OT, PT, speech-language therapies
- Home care, creative companion care, residential options

Advocate for benefits

- Retirement and ST, LT disability benefits
- Compassionate Allowances, SSDI, Medicare

Facilitate coordination of care

- Transitions common and challenging
- Acute hospitalizations & med changes

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FTD Research

Momentum in FTD science is growing

Biomarkers research

- Improve diagnosis

Drug development & Clinical Trials

- Different mechanisms needed for different biological subtypes
- Tau, TDP43, GRN mutation carriers, C9ORF72 mutation carriers

Care and services research

- Applied technology to improve care

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FTD Research

Participation is critical



www.ftdregistry.org

3006 PERSONS JOINED THE REGISTRY	1207 FAMILY MEMBERS
575 PERSONS DIAGNOSED WITH FTD	1224 CAREGIVERS

[JOIN THE REGISTRY](#)

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AFTD Provides Help & Hope

AFTD's mission is to improve the quality of life of people affected by FTD and drive research to a cure.

Support | Education | Research | Advocacy | Awareness

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AFTD Resources



www.theAFTD.org

HelpLine: 866-507-7222
info@theAFTD.org

www.AFTDkidsandteens.org

Support Services



2306
HelpLine cases
in 2018

100

AFTD-affiliated
support group
leaders in 32 states

8

AFTD-affiliated
phone/internet
groups

277

Comstock grants
awarded (148
respite, 46 travel,
20 quality of life)



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Partners in FTD Care

**In-depth education on care challenges
Case-stories, concise strategies & tips**

- Challenges in Diagnosis
- ALS / FTD
- Managing Apathy
- Anosognosia
- Medications in FTD
-And more



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theaftd.org | HelpLine: 1-866-507-7222 | info@theaftd.org
