Introduction to FTD: Advancing Help and Hope

Kansas Alzheimer’s Association Education Conference
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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.

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
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.

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:

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
FTD vs Alzheimer’s Disease

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<thead>
<tr>
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<th>FTD</th>
<th>Alzheimer’s</th>
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<tbody>
<tr>
<td><strong>Age of onset</strong></td>
<td>Av. 50’s-60’s</td>
<td>&gt; 65; av. ~ 80</td>
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<tr>
<td><strong>Prevalence in US</strong></td>
<td>50,000-60,000</td>
<td>5.8 million</td>
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<td><strong>Clinical hallmarks</strong></td>
<td>Behavior, language, movement</td>
<td>Memory loss</td>
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<tr>
<td><strong>% Inherited</strong></td>
<td>10-20%</td>
<td>&lt; 1%</td>
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<td><strong>Time to diagnosis</strong></td>
<td>3.6 years</td>
<td>2.8 years</td>
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FTD Syndromes and Diagnosis

FTD is a Spectrum of Diseases
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

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

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
What you may see:

- **Compulsive behaviors** - verbal or motor eg: hand rubbing, clapping, counting aloud, humming, catch phrases
- **Complex repetitive motor routines** - walking a fixed route, collecting & hoarding objects, counting money, checking and rituals involving unusual toileting and hygiene behavior
- **Eating changes** - Narrow food preferences, eg: eating only ice cream; eating to excess; taking food off another's plate
- **Loss of self-awareness** or denial of symptoms (anosognosia)
  No recognition of behavior changes or their effect

Primary Progressive Aphasia (PPA)

**Early and progressive change in language skills**

Memory, self-awareness relatively preserved

**Nonfluent** – Agrammatic PPA
- Problems in expressive language

**Semantic PPA**
- Loss of word meaning

**Logopenic PPA**
- Impaired word retrieval

Movement Disorders

**Progressive Supranuclear Palsy (PSP)**
- Balance, unexplained falls
- Slow stiff movements
- Trouble coordinating eye movements

**Corticobasal Syndrome (CBS)**
- Slow, reduced movements
- Rigidity
- Apraxia
- Limb and fine motor control issues
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

"That's when I knew something was seriously wrong"

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.

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

Impact on Family System
Feeling isolated magnifies challenges
- Roles change-
  - Young adult and school age children
  - Behaviors intrusive, embarrassing
  - Friends pull away
  - Ambiguous loss
Economic Burden Study*

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.

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 (e.g., 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

- Awareness of Research
- Individualized & Coordinated Care
- Disease Education
- Help & Hope
- FTD Peer Support
- 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, ICFD 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

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**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

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

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
FTD Research
Participation is critical

ALLFTD – NIH Grant
• 19 centers US & Canada
• Natural history studies
• Clinical trial network

FTD Disorders Registry
• Contact and research registry
• Persons diagnosed, family, care partners

www.ftdregistry.org

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

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 50 states
- 8 AFTD-affiliated phone/internet groups
- 277 Comstock grants awarded (49 respite, 46 travel, 20 quality of life)

Partners in FTD Care

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

- Challenges in Diagnosis
- ALS / FTD
- Managing Apathy
- Anosognoisa
- Medications in FTD

...And more

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