



**VA** GREATER LOS ANGELES  
HEALTHCARE SYSTEM



*A Division of VA Desert Pacific  
Healthcare Network*

# **Frontotemporal Degeneration: An Update**

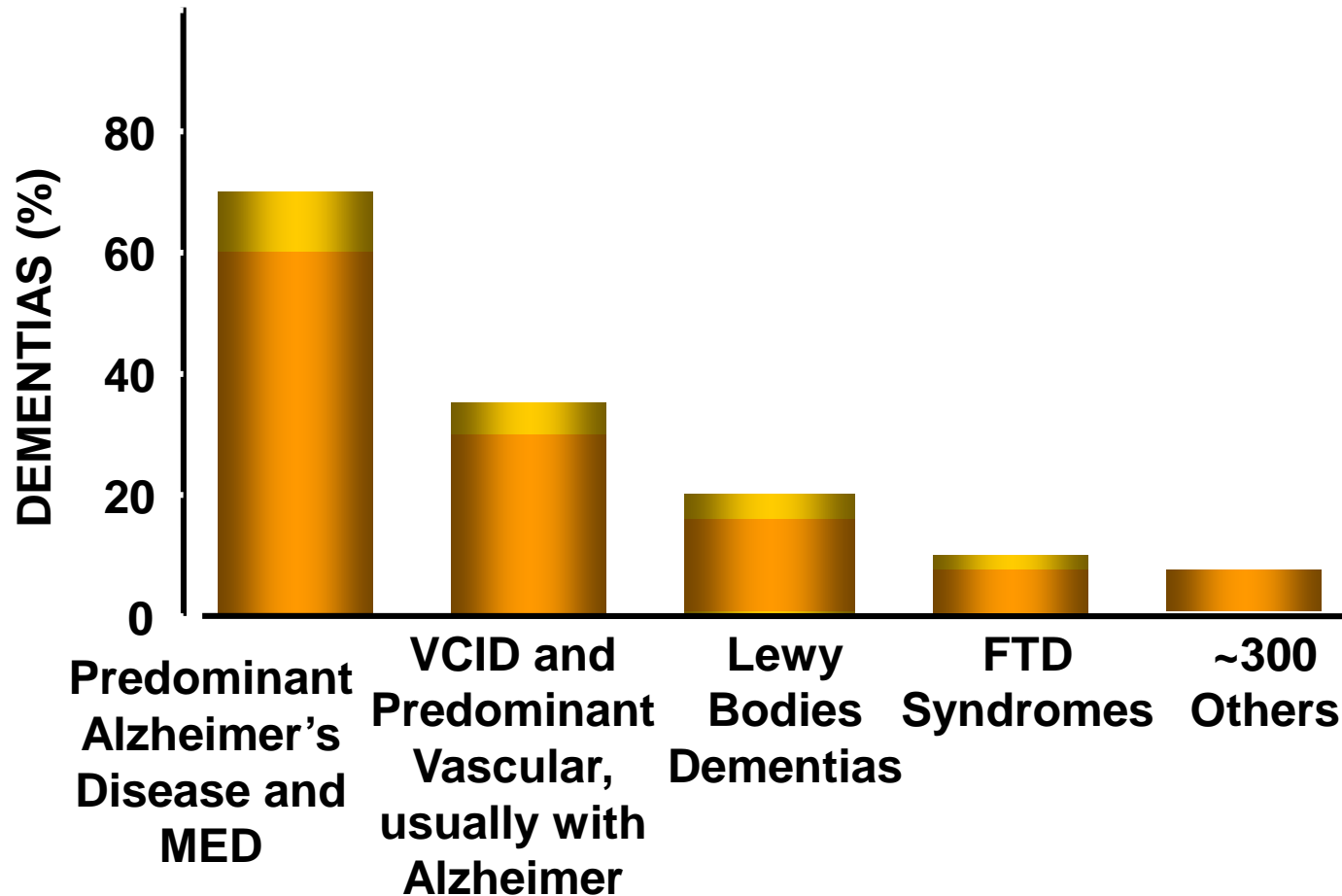
## **AFTD Educational Conference Los Angeles 2019**

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# PART 1: FTD Syndromes



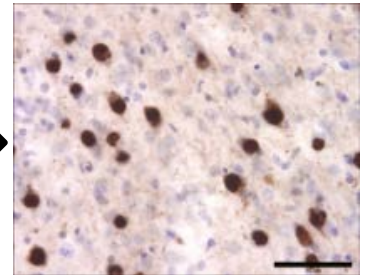


# Brief History of FTD

**1892** Arnold Pick described first FTD patient

**1911** Alzheimer described pathology:

Silver-stained Pick bodies →



## **DARK AGES**

**1993+** *Renaissance*: Epidemiology, Clinical Criteria

**1997** *Age of Tauopathy*: abn. *tau*, FTDP-17, MAPT gene

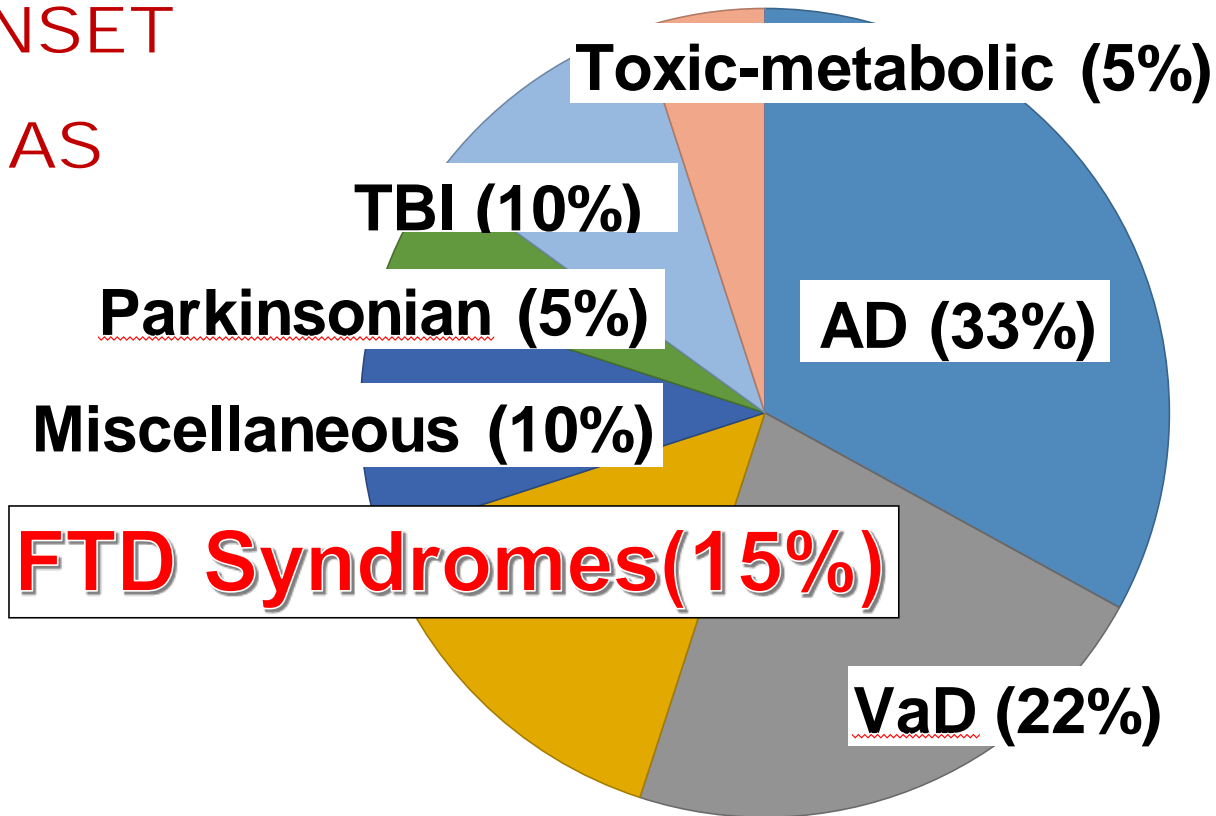
**2006** *Age of TDP-43 and progranulin gene*

**2008-9** *Expanded genetics and path: TARDBP gene, FUS*

**2011** *C'9orf72 and interface with ALS and psychosis*

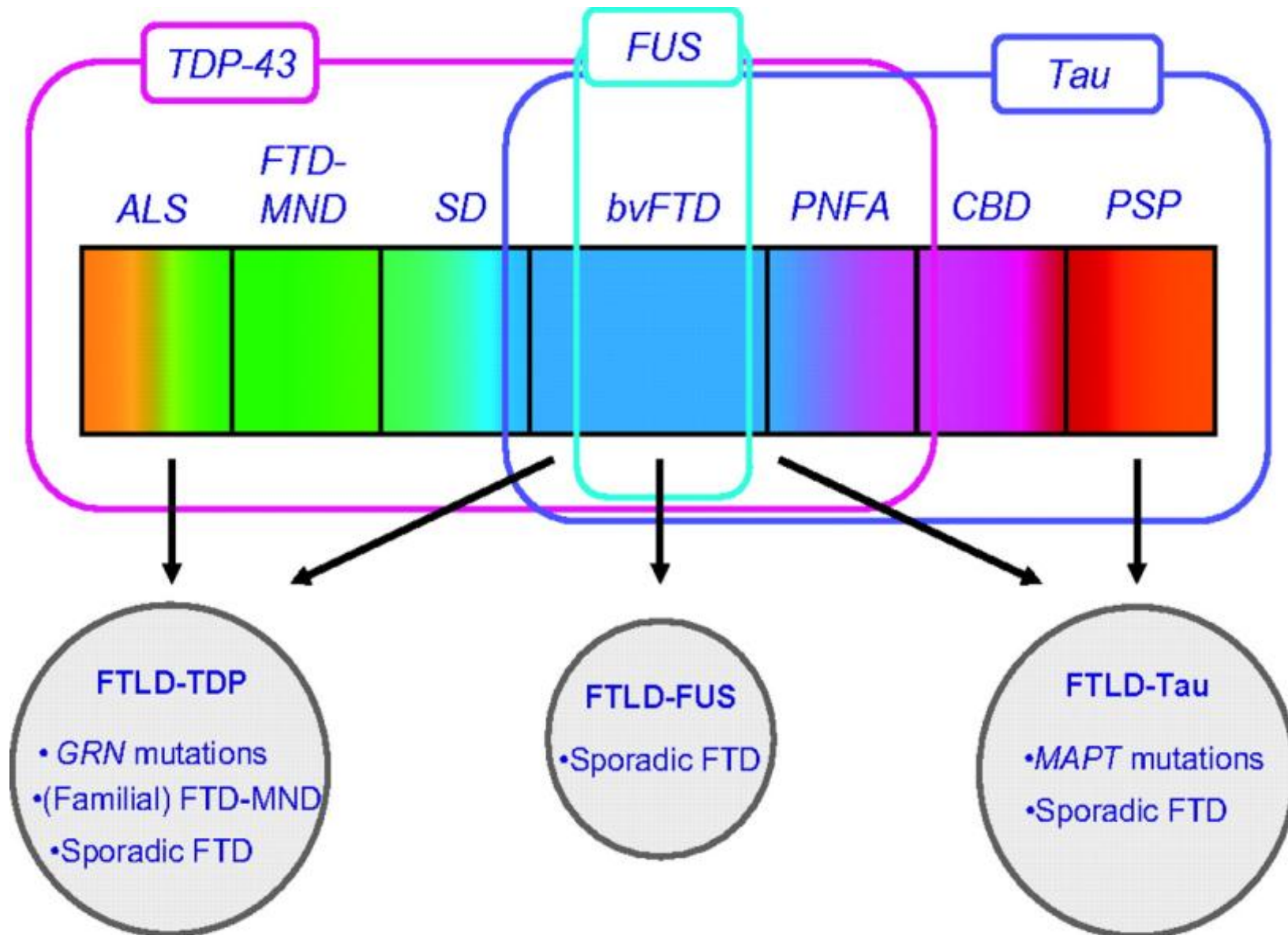
**2015** Multicenter funding for clinical trials in FTD

EARLY-ONSET  
DEMENTIAS



1. ~8 (5-15)% of dementias or ~15% if onset is <65
2. FTD:AD-~1:65-70 overall or 1:2-3 if <65; 1:1 if <60
3. Onset 57-58 (20-80+); no significant sex difference
4. BvFTD duration 7.6yrs (3-10), longer for PPA, SD

# NEURODEGENERATIVE SPECTRUM OF FTD-RELATED SYNDROMES

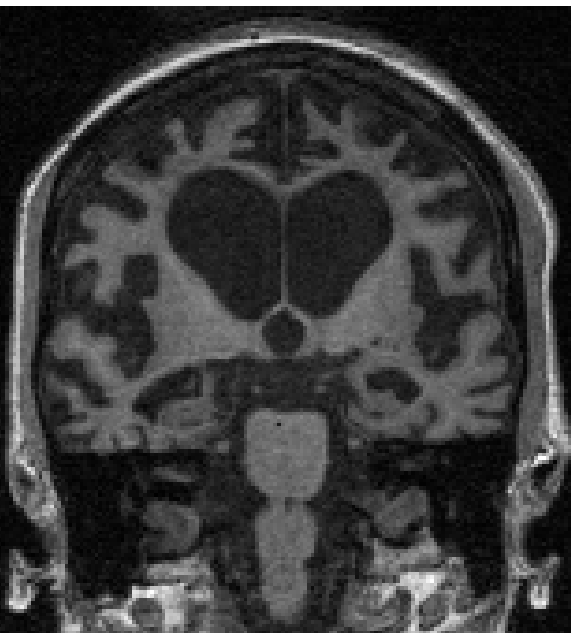
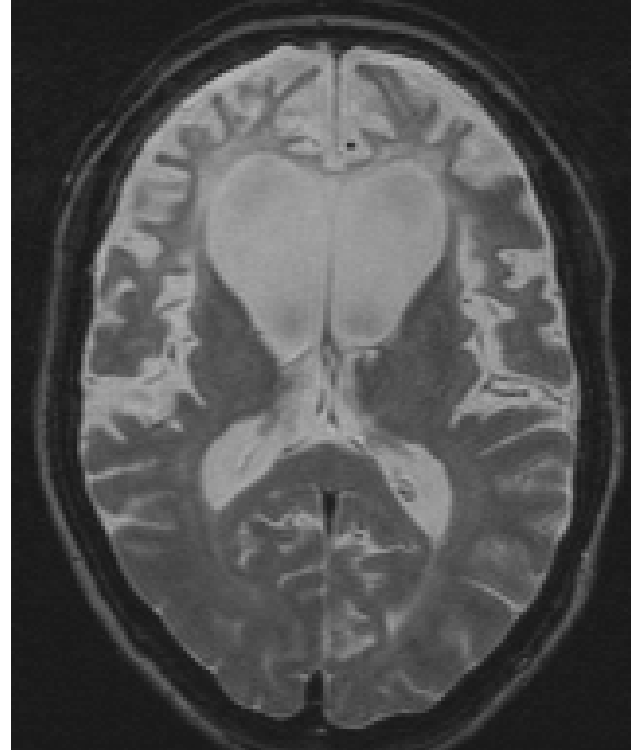
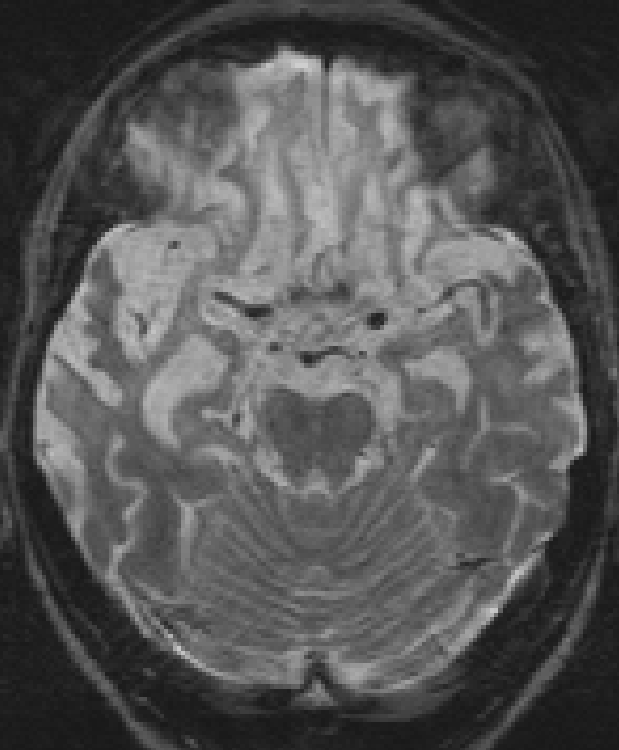


# **Intl Consensus Criteria for bvFTD**

**Neurodegenerative Disease (progressive)**

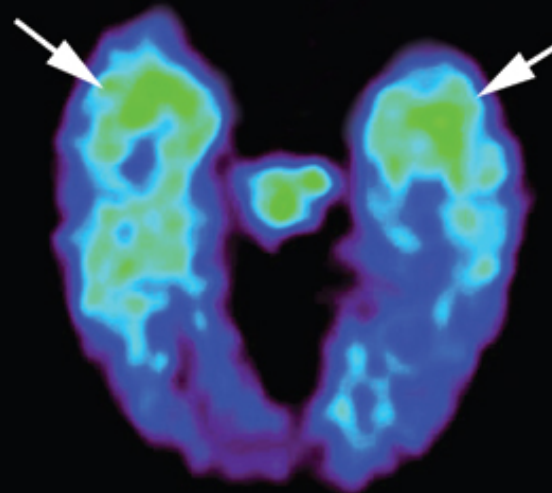
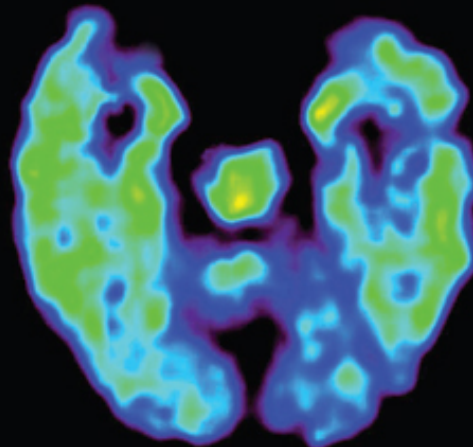
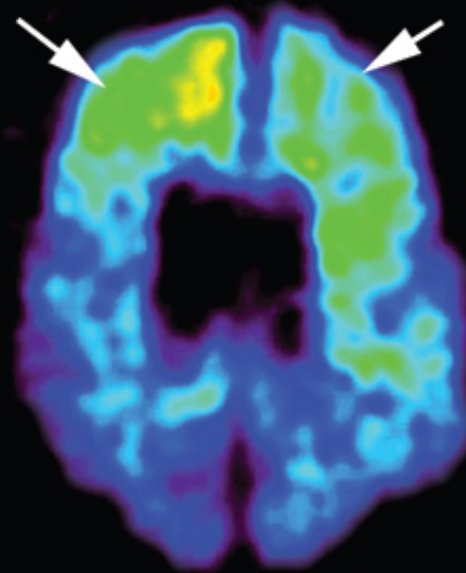
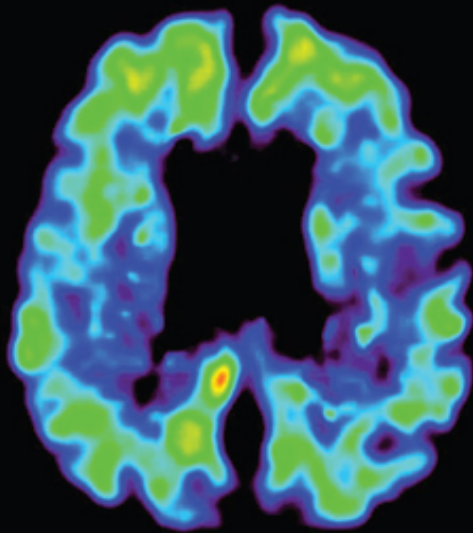
**Possible bvFTD (3 of A-F present) (“Probable” if additional neuroimaging)**

- A. Early behavioral disinhibition**
- B. Early apathy or inertia**
- C. Early loss of sympathy or empathy**
- D. Early perseverative, stereotyped or compulsive/ritualistic behavior**
- E. Hyperorality and dietary changes**
- F. Neuropsychological profile (all 3 present)**



**ALZHEIMER'S  
DISEASE**

**FRONTAL LOBE  
DEMENTIA**



**[F-18]FDDNP PET**



# Neuropathology and Genetics

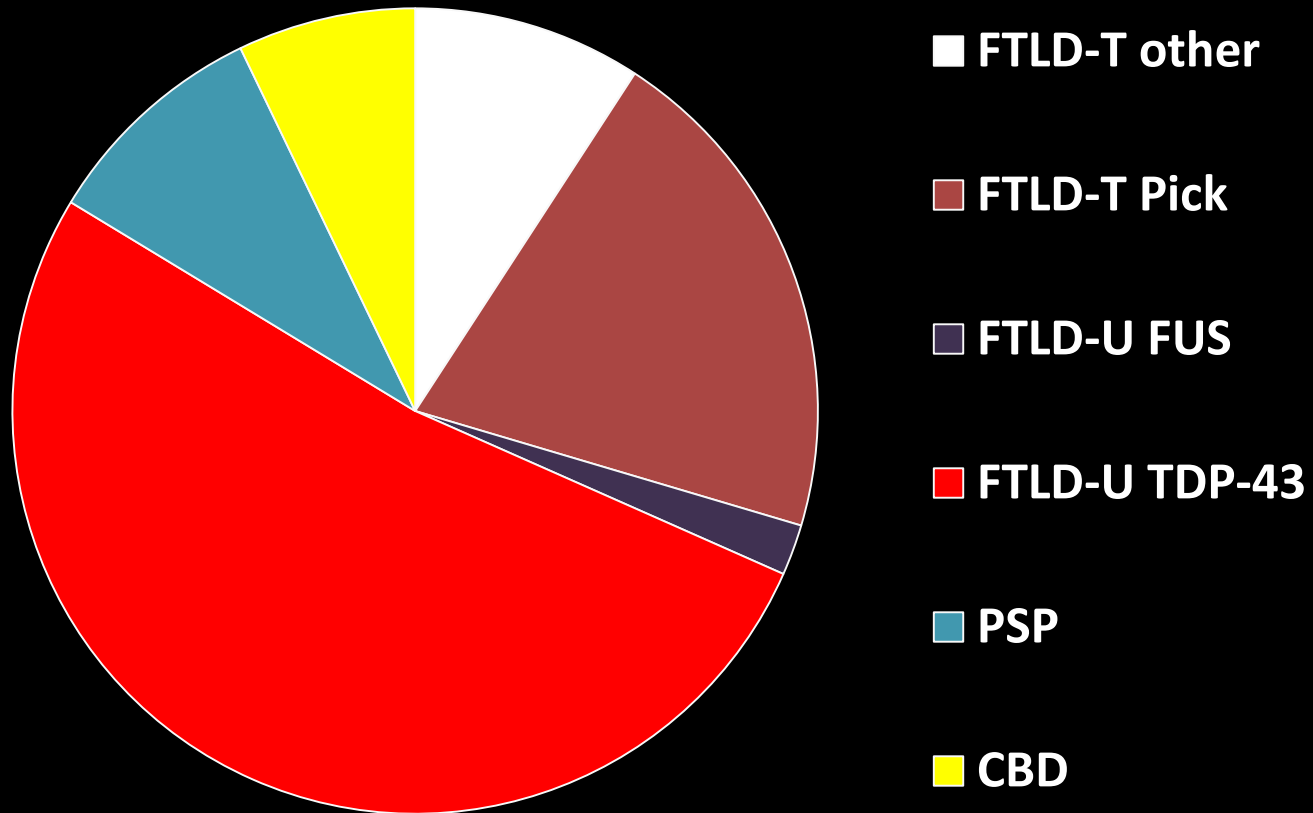
## **Variable Neuropathology termed “Frontotemporal Lobar Degeneration” (FTLD):**

All have frontotemporal regional neuronal loss, microvacuolarization, and neuronal inclusion bodies with abnormal protein deposits -primarily hyperphosphorylated tau or transactive response DNA-binding protein tau k43 (TDP-43)

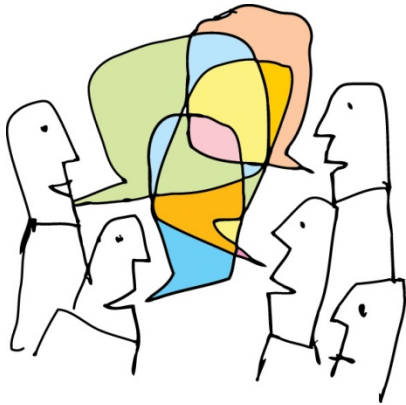
## **Variable Genetics: 33% or more have suggestive FHx**

13% caused by an autosomal dominant gene – C'9orf72, PRGN, MAPT, VCP, CHMP2B, TARDBP

# FTLD Neuropathology



# BvFTD is characterized by Altered Social Behavior



- Altered social behavior affects the psychological well-being and social life of families and caregivers.
- Understanding altered social behavior is critical for behavior management
- Accommodating the behavior in a calm, safe environment while providing education and support for the caregiver, is more important than extinguishing the behavior



# What is the impact on patients?

- Loss of independence
- Loss of role outside home (eg, occupation)
- Loss of role in family
- Social isolation and exclusion
- Decreased overall sense of self/identity

# What is the impact on families?

- Altered balance between patient needs and family needs
- Altered family roles
- Ambiguity about the future and how to plan
- Family resilience and tensions about caregiving
- Children—discussing and helping them cope

# Major Social Behavior Disturbances in BvFTD

1. Detachment: unmotivated, apathetic, “inertia”
2. Disinhibition: violate social norms/manners
3. Altered interpersonal connection or loss of empathy
4. Altered communication

# Disengagement or Apathy

**Apathy has the most impact on marital relationship** (du Vugt et al., 2003)

- Provide structure - more effective than free time
- Offer or direct to individual or small group activities
- Do not force them; let them passively participate
- Ensure tasks are simple so that they can complete
- At onset, explain activities in simple language

# Disinhibition

**In Germany, behavioral disturbances were predominant reason for hospital admission among 58 patient with FTLD included FTD** (Ibach et al, Dement Geriatr Cogn disord 2004;17:269-73)

- Identify trigger for disinhibition and interventions
- Avoid confrontation; gently redirect to another activity
- Reduce environmental stimulation
- Involve other family members and caregivers
- If disruptive, inform others, include what does or does not work



# Altered Interpersonal Connection

**FTD caregivers report a loss of emotional attachment leading to isolation and anger due to behavioral symptoms** (Massimo et al, Geriatr Nurs 2013;34:302-6)

- Rethink expectation of emotional feedback; offer empathy without expecting reciprocity
- Provide them information about others' perspectives
- Encourage families to share what they did together
- Share moments of connection and special events
- Instruct others so they don't expect validation

# Altered Communication

**Many have little verbal output and single or short phrase answers and others are excessively talkative and jocular.**

- Approach with calm, patient, pleasant tone of voice
- Reduce competing stimulation and distractions
- Use the same terms consistently for care issues
- Other forms of communication: touch and lead, hand motions, props, picture, sing, short written words
- Technology—iPads with communication apps and software programs like Proloquo2Go([www.proloquo2go.com](http://www.proloquo2go.com))



# Caregiver

**Worse strain, emotional distress and lower perceived control among bvFTD caregivers. Levels of depression for FTD caregivers are twice that of AD caregivers** (Wong et al, 2012 AMP 20;724-8) (Moishi et al, Dement Geriatr Cogn disord 2009;27:76-81).

- Practice caregiver wellness, self-care, and forgiveness
- Find balance: spend time together AND time apart
- Have realistic expectations
- Reach out and talk to others about what is happening
- Support groups with other caregivers of those with FTD

**I hope that this information helps you.  
Thank you very much for your attention.**

