

# The new NACC FTLD Module

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Workgroups

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# Purpose of FTLD Module

- To capture salient information about the FTLD syndromes not currently available in AD-oriented current UDS
  - No changes in current UDS “allowed”
- To have the FTLD module mesh with the current UDS
- To foster collaborative, multicenter research in the FTLD’s

# FTLD Collaboration

- Instigated by Tony Phelps at NIA in conjunction with Walter Koroshetz at NINDS
- Data forms and database designed by Bud Kukull, Duane Beekly and Elizabeth Robichaud
- Core Leaders
  - Clinical: Knopman, Mendez, Grossman, Mesulam
  - Neuropsychology: Miller, Kramer, Weintraub, Hillis, Grafman, Rankin
  - Genetics: Boeve, Rademakers, Mayeux, Geschwind, Shellenberg, Bird
  - Imaging: Rosen, Dickerson, Josephs, Gee, Foster

# FTLD Clinical Module

- What is available in current UDS
  - Augmented (8-domain) CDR
  - Neuropsychiatric inventory
  - UPDRS
  - Diagnostic choices for
    - bvFTD
    - Nonfluent variant PPA
    - Semantic variant PPA
    - Logopenic variant PPA
    - Progressive Supranuclear Palsy
    - Corticobasal Syndrome

# FTLD Clinical Module

- What is missing in UDS
  - All detail on basis for diagnoses on:
    - Aphasia
    - Specific behaviors typically abnormal in bVFTD
    - Aspects of motor exam not covered by our current UPDRS

# FTLD Clinical Module

- In the new FTLD module
  - Augmented neurological examination
  - Specific features in aphasia assessment for PPAs
  - Specific features for making diagnosis of bvFTD



**B. Is there evidence for corticobasal syndrome?**      No  0 *If no, go to section C.*  
 Yes  1 *If yes, complete section B, below.*

	Absent	Present	Unstable (specify reason)
<b>B1. LIMB APRAXIA</b>			
a. Right upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
b. Left upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
<b>B2. ALIEN LIMB PHENOMENON</b>			
a. Right upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
b. Left upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
<b>B3. CORTICAL SENSORY LOSS</b>			
a. Right upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
b. Left upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
<b>B4. DYSTONIA</b>			
a. Right upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
b. Left upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
c. Right lower extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
d. Left lower extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
<b>B5. MYOCLONUS</b>			
a. Right upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____
b. Left upper extremity	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9 _____





**INITIAL VISIT PACKET** NACC UNIFORM DATA SET (UDS) — FTD MODULE

**Form B9F: Clinical PPA and bvFTD Features**

Center: \_\_\_\_\_ Subject ID: \_\_\_\_\_ Form Date: \_\_\_\_/\_\_\_\_/\_\_\_\_

**NOTE:** This form is to be completed by a clinician with experience in evaluating patients with frontotemporal lobar degeneration. For additional clarification and examples, see FTD Coding Guidebook for Initial Visit Packet, Form B9F. Check only one box per question.

Visit #: \_\_\_\_\_

Examiner's initials: \_\_\_\_\_

*Initiate and complete this form, based on how subject presents at this visit, only if you believe that the subject's underlying disorder is of a neurodegenerative type.*

**Clinical PPA: Speech and language symptoms (Questions 1 – 13)**

	No	Yes
1. <b>Does patient have clinically important alterations in speech and language?</b> Are there disturbances of expressive speech, other than simple dysarthria, word-finding difficulties, or difficulties with comprehension of spoken language?	<input type="checkbox"/> 0	<input type="checkbox"/> 1
2. Are these alterations in speech and language the principal cause of impaired daily living activities?	<input type="checkbox"/> 0	<input type="checkbox"/> 1
3. Are/were the alterations in speech and language the most prominent deficit during the first initial stages (1–2 years) of the disorder?	<input type="checkbox"/> 0	<input type="checkbox"/> 1

*If clinically important alterations in speech and language are present, please describe their occurrence and severity below. The symptoms listed here are part of the core diagnosis for PPA subtypes.*

	Absent	Questionably present	Present but mild	Present moderate	Present severe
4. Poor object naming	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
5. Impoverished word selection/retrieval in spontaneous speech	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
6. Grammatical simplification or grammatical errors in speech	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
7. Effortful, halting speech	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
8. Speech sound / word errors (paraphasias)	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
9. Impaired single-word comprehension	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
10. Poor object / person knowledge	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
11. Circumlocutory speech, empty speech	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
12. Surface dyslexia	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
13. Impaired speech repetition	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4

bvFTD features: Behavioral symptoms (Questions 14 – 21)

	No	Yes
14. Does patient have clinically important alterations in behavior, personality, or comporment consistent with bvFTD (root symptom of bvFTD)?	<input type="checkbox"/> 0	<input type="checkbox"/> 1
15. Are these alterations in behavior, personality, or comporment the principal cause of impaired daily living activities	<input type="checkbox"/> 0	<input type="checkbox"/> 1
16. Were/are the alterations in behavior, personality, and comporment the most prominent deficit during the first initial stages (1–2 years) of the disorder.	<input type="checkbox"/> 0	<input type="checkbox"/> 1

*If clinically important alterations in behavior are present, please complete the following checklist.*

	Absent	Questionably present	Present but mild	Present moderate	Present severe
17. <b>Disinhibition</b> Socially inappropriate behavior; loss of manners or decorum; impulsive, rash, or careless actions	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
18. <b>Apathy</b> Loss of interest, drive, and motivation; decreased initiation of behavior	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
19. <b>Loss of sympathy / empathy</b> Diminished response to to other people's needs or feelings; diminished social interest, interrelatedness, or personal warmth	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
20. <b>Ritualistic / compulsive behavior</b> Simple repetitive movements or complex compulsive or ritualistic behaviors	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
21. <b>Hyperorality and appetite changes</b> Altered food preferences, binge eating, increased consumption of alcohol or cigarettes, oral exploration or consumption of inedible objects	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4

# FTLD Neuropsychological Module

- What is available in current UDS that is relevant to FTLD
  - Fluency
  - Digit Span
  - Boston Naming
  - Trailmaking

# FTLD Neuropsychological Module

- What is missing in current UDS
  - Executive functioning
  - Language testing
    - Sentence repetition
    - Grammatical knowledge
    - Semantic fluency
    - Surface dyslexia
  - Social cognition

# FTLD Neuropsychological Module

- **In the new FTLD module**
  - “Flanker” a test of attention and response inhibition
  - Additional semantic and letter fluency tests
  - Picture vocabulary
  - NW anagram test, a test of grammatic knowledge
  - Sentence repetition
  - Questionnaires on social behavior, social norms, dynamic affect recognition, insight

# FTLD Genetics Module

- What is available in current UDS that is relevant to FTLD
  - Asks only about “dementia yes/no”
- What is needed for FTLD
  - Specific dementia diagnoses and method of ascertainment of dx (eg autopsy, family report, record review, etc)
- Note: information on mutation carrier status not recorded



**INITIAL VISIT PACKET** NACC UNIFORM DATA SET (UDS) — FTD MODULE — GENETICS

# Form A3F: Family History Supplement

Center: \_\_\_\_\_ Subject ID: \_\_\_\_\_ Form Date: \_\_\_\_/\_\_\_\_/\_\_\_\_

**NOTE:** This form is to be completed by a clinician with experience in evaluating patients with frontotemporal lobar degeneration. For additional clarification and examples, see FTD Coding Guidebook for Initial Visit Packet, Form A3F.

Visit #: \_\_\_\_\_

Examiner's initials: \_\_\_\_\_

PARENTS		
	a. Method*	b. Primary DX**
1. Mother	_____	_____
2. Father	_____	_____

**\*Codes for Method of Evaluation**  
See page 3 of form for descriptions

- 1 Autopsy
- 2 Examination
- 3 Medical record review from formal dementia evaluation
- 4 Review of general medical records AND informant and/or subject telephone interview
- 5 Review of general medical records only
- 6 Subject and/or informant telephone interview
- 7 Family report

**AFFECTED SIBLINGS** — Use the form below to provide information on affected siblings only. Sibling Number on this form comes from UDS Initial Visit Form A3, which lists ALL siblings. The first-born sibling is Sibling Number S01, the third-born sibling is S03, the n<sup>th</sup>-born sibling is S0n, etc. Please check birth dates on UDS Initial Visit Form A3 to insure correct birth-order numbering on this form.



# FTLD Imaging Module

- What is available in current UDS that is relevant to FTLD
  - Whether imaging has been performed
  - Whether there is evidence for cerebrovascular disease
- What is needed for FTLD
  - More detail on imaging abnormalities in MR, FDG-PET

# FTLD and Neuropathology

- What is available in current UDS that is relevant to FTLD
  - Core parts of UDS neuropath readily accommodate FTLD except...
  - No items for TDP-43 subtyping
  - No FUS items
- But, it doesn't make sense to have a new module that is FTLD specific for neuropath as this material should be in standard UDS, and therefore will have to await new version

# So how will the FTLD module work?

- Voluntary participation by ADC's!!
- To be completed in conjunction with standard UDS in patients with syndromes of bvFTD, PPA's (and selected control normals, AD cases)
- Module use needs to be anticipated because extra time required for neuropsych and informant interview

# Additional time needed for FTLD module

- Clinical by examining MD : ~ 10 minutes
- Caregiver interviews: ~ 20-30 minutes
- Family history interview: negligible
- Neuropsychological testing: ~ 45 minutes
- Additional Form completions: ??

# Next Steps

- Pilot Test module in selected centers
- Training session for new module
- Make fully operational
- Obtain funding for multicenter FTLD research
  - Risk factors
  - Genetic studies
  - Imaging
  - Clinical trial instrument development