The new NACC FTLD Module

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And thanks to NACC, especially Elizabeth Robichaud et al

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 Clinical Module

- In the new FTLD module
  - Augmented neurological examination
  - Specific features in aphasia assessment for PPAs
  - Specific features for making diagnosis of bvFTD
  - Neuropsychological battery
  - Improved genetic info form
  - More Imaging information

Supplementary Neurologic Examination

Motor neuron disease

Eye mvmt Disorder

Corticobasal syndrome

	Not to a degree that would justify such a diagnosis[1]	Yes — L>R[2]	Yes — R>L[3]	Yes — without major asymmetry[4]
SECTION A				
A1. Does the patient have limb or torso fasciculations consistent with a diagnosis of spinal muscular atrophy (SMA)[5] or amyotrophic lateral sclerosis (ALS)[6]?	☐ 0	☐ 1	☐ 2	☐ 3
A2. Does the patient have limb weakness and/or hyperreflexia consistent with a diagnosis of primary lateral sclerosis (PLS)[7] or ALS[6]?	☐ 0	☐ 1	☐ 2	☐ 3
A3. Does the patient have bulbar weakness and/or fasciculations consistent with a diagnosis of ALS[6]?	☐ 0			☐ 3
A4. Does the patient have eye movement abnormalities consistent with a diagnosis of progressive supranuclear palsy (PSP)[8]?	☐ 0			☐ 3
A5. Does the patient have dystonia or other motor signs consistent with a diagnosis of corticobasal degeneration (CBD)[9]?	☐ 0	☐ 1	☐ 2	☐ 3
A6. Is there ideomotor apraxia[10]?	☐ 0	☐ 1	☐ 2	☐ 3
A7. Is the alien limb phenomenon[11] present?	☐ 0	☐ 1	☐ 2	☐ 3
A8. Is there myoclonus[12]?	☐ 0	☐ 1	☐ 2	☐ 3

Supplementary Neurologic Examination:
Generalizable Gait Assessment

SECTION B Gait disturbances	
B1. Severity	☐ 0 Normal ☐ 1 Slight alteration in speed or fluidity of gait ☐ 2 Walks with difficulty but requires no assistance ☐ 3 Severe disturbance ☐ 4 Cannot walk at all ☐ 8 Untestable (specify reason): _____
B2. Type	☐ 0 Normal ☐ 1 Hemiparetic (spastic) ☐ 2 Foot drop gait (lower motor neuron) ☐ 3 Ataxic gait ☐ 4 Other non-parkinsonism gait (axial rigidity, unstable) ☐ 5 Parkinsonian gait (stooped, shuffling, reduced arm swing) ☐ 8 Untestable (specify reason): _____

PPA: Gateway Questions:

Clinical PPA: Speech and language symptoms (Questions 1 – 13)			
	No	Yes	Not evaluated
1. **Does patient have clinically important alterations in speech and language?** Other than simple dysarthria, are there difficulties with word-finding, comprehension of spoken language, or disturbances of expressive speech? *If "No," skip to Item 14.*	☐ 0	☐ 1	☐ 9
2. **Are these alterations in speech and language the principal cause of impaired daily living activities?**	☐ 0	☐ 1	☐ 9
3. **Are/were the alterations in speech and language the most prominent deficit during the first initial stages (1–2 years) of the disorder?**	☐ 0	☐ 1	☐ 9

PPA: Specific Items

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

Matches Gorno-Tempini Neurology formulation

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bvFTD Gateway Questions

bvFTD: Behavioral symptoms (Questions 14 – 21)			
	No	Yes	Not evaluated
14. Does patient have clinically important alterations in behavior, personality, or comportment consistent with bvFTD (root symptom of bvFTD)? *If answer is "No" or "Not Evaluated," END FORM HERE.*	☐ 0	☐ 1	☐ 9
15. Are these alterations in behavior, personality, or comportment the principal cause of impaired daily living activities?	☐ 0	☐ 1	☐ 9
16. Were / are the alterations in behavior, personality, and comportment the most prominent deficit during the first initial stages (1 – 2 years) of the disorder?	☐ 0	☐ 1	☐ 9

bvFTD Specific Diagnostic Items

If clinically important alterations in behavior are present, please complete the following checklist.	Absent	Questionably present	Present but mild	Present moderate	Present severe	Not evaluated
17. **Disinhibition** Socially inappropriate behavior; loss of manners or decorum; impulsive, rash, or careless actions	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 9
18. **Apathy** Loss of interest, drive, and motivation; decreased initiation of behavior	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 9
19. **Loss of sympathy / empathy** Diminished response to to other people's needs or feelings; diminished social interest, interrelatedness, or personal warmth	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 9
20. **Ritualistic / compulsive behavior** Simple repetitive movements or complex compulsive or ritualistic behaviors	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 9
21. **Hyperorality and appetite changes** Altered food preferences, binge eating, increased consumption of alcohol or cigarettes, oral exploration or consumption of inedible objects	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 9

Matches proposed criteria from CFTD

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FTLD Neuropsychological Module in Pilot

- “Flanker” a test of attention and response inhibition
- Additional semantic and letter fluency tests
- Semantic association tests – 16 items
- Noun and verb naming – 30 items
- Visual memory – Benson figure
- NW anagram test, a test of grammatic knowledge – 10 items
- Word repetition 20 items
- Sentence repetition and reading – 5 items
- Behavioral Inhibition Scale - 7 items
- Interpersonal Reactivity index -14 items
- Self Monitoring - 13 items
- Dynamic affect recognition -12 items
- Behavior observations – 14 items
- Social Norms - 22 items

FTLD Neuropsychological Module

- **Piloted version seems to be too long**
- **Need to reconsider what the neuropsychology battery should do in light of NACC philosophy**
  - Brief but informative
  - Shorter forms of language tests needed, but are not validated
  - Shorter forms of behavior questionnaires also not validated

Genetics Module: more general than current A3

Form A3F: Family History: Affected Family Members

Center: _____ Subject ID: _____ Form Date: ____/____/_____

Visit #: ____

Examiner's initials: ____

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.

Any neurological diagnosis

"AFFECTED FAMILY MEMBERS" — For the purposes of Form A3F, "affected" means affected by dementia **OR** by any of the non-normal clinical diagnoses listed in Appendix 1 on page 4 of this form.

AFFECTED FAMILY MEMBERS	
1. Are there affected family members? (See box above for definition of "affected.") **If the answer is "No" or "Unknown," please skip the rest of this form.**	☐ 0 No ☐ 1 Yes ☐ 9 Unknown

AFFECTED PARENTS — Use the form below to provide information on affected parents only (see definition of "affected" in the box above).

AFFECTED PARENTS	a. Neurological problem*	b. Primary DX**	c. Method***	d. Age of onset
2a. Mother	___	_____	___	___ ___
2b. Father	___	_____	___	___ ___

***Codes for neurological problem**

0 No neurologic or psychiatric disorder or diagnosis
1 Cognitive impairment/behavior change
2 Parkinsonism
3 ALS
4 Other neurologic condition such as multiple sclerosis or stroke
5 Psychiatric condition such as schizophrenia, bipolar disorder, or alcoholism
6 Other condition
9 Unknown

****Codes for primary diagnosis**

See Appendix 1 on page 4 of this form

*****Codes for**

For descriptions, see Appendix 2 on page 4 of this form

1 Autopsy
2 Examination
3 Medical record review from formal dementia evaluation

Method of Dx needed

NACC UNIFORM DATA SET (UDS) – FTD MODULE

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

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

Next Steps

- Re-design neuropsych component
- Make fully operational
- Obtain funding for multicenter FTLD research
  - Risk factors
  - Genetic studies
  - Imaging
  - Clinical trial instrument development