

INITIAL VISIT PACKET NACC UNIFORM DATA SET (UDS) — FTLD 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 subjects with frontotemporal lobar degeneration. For additional clarification, see FTLD Coding Guidebook for Initial Visit Packet, Form B9F. Check only one box per question.

Visit #: _____
 Examiner's initials: _____

Gateway question for primary progressive aphasia (PPA)		
	No	Yes
<p>1. Does the subject have an acquired and progressive difficulty with language* consistent with PPA of a neurodegenerative type?</p> <p>*DIFFICULTY WITH LANGUAGE: Other than simple dysarthria, are there difficulties with retrieving, using, repeating, sequencing, or understanding words?</p> <p>If answer is "No", check "0 (Absent)" for Questions 2–11 and "0 (No)" for Question 12.</p>	<input type="checkbox"/> 0	<input type="checkbox"/> 1

Characterizing speech and language symptoms / assigning PPA subtype				
<i>Are these features present on the current examination? Note: many of these items are also evaluated in the neuropsychological assessment. The responses recorded here should represent the consensus of the clinical and neuropsychological evaluation.</i>	Absent	Questionably present	Definitely present	Not evaluated
<p>2. Poor object naming (Core diagnostic feature of semantic variant; abnormal in all variants)</p>	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
<p>3. Impoverished word selection / retrieval in spontaneous speech or writing (Core diagnostic feature of logopenic variant; abnormal in all variants)</p>	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
<p>4. Impaired word comprehension (Core diagnostic feature of semantic variant; absent in other variants)</p>	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
<p>5. Poor object/person knowledge (Secondary diagnostic feature of semantic variant; absent in other variants)</p>	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
<p>6. Grammatical simplification or grammatical errors in speech or writing (Core diagnostic feature of nonfluent/agrammatic variant)</p>	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
<p>7. Effortful, halting speech (Core diagnostic feature of nonfluent/agrammatic variant)</p>	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9

Characterizing speech and language symptoms / assigning PPA subtype (continued)				
	Absent	Questionably present	Definitely present	Not evaluated
8. Circumlocutory, empty speech (Secondary diagnostic feature of logopenic variant; also present in semantic variant)	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
9. Speech sound/word errors (paraphasias) (Secondary diagnostic feature of logopenic variant; abnormal in nonfluent/agrammatic variant)	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
10. Impaired speech repetition (inability to repeat verbatim sentence-length material) (Core diagnostic feature of logopenic variant; present in nonfluent/agrammatic type; absent in semantic variant)	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
11. Surface dyslexia and dysgraphia — <i>also refer to Word Reading Test from FTLN Neuropsychological Battery</i> (Secondary feature of semantic variant)	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
12. ROOT DIAGNOSIS OF PPA Does the subject have an acquired and progressive difficulty with language consistent with PPA of a neurodegenerative type AND is the language disorder the most prominent deficit at symptom outset and for the initial phase (1–2 years) of the disorder?	<input type="checkbox"/> 0 No Proceed to Question 14		<input type="checkbox"/> 1 Yes — Meets root diagnosis of PPA Proceed to Question 13	
13. Consensus diagnosis of dominant PPA subtype based on clinician and neuropsychologist judgment <i>NOTE: The diagnostic criteria in this module do not match the criteria in UDS V2.0 (Form D1). While Version 2.0 of the UDS is still in use, keep the two sets of diagnostic criteria separate.</i>	<input type="checkbox"/> 1 PPA, semantic variant (semPPA) <input type="checkbox"/> 2 PPA, nonfluent/agrammatic variant (nf/gPPA) <input type="checkbox"/> 3 PPA, logopenic variant <input type="checkbox"/> 4 PPA not otherwise specified			

Gateway question for behavioral variant frontotemporal dementia (bvFTD)		
	No	Yes
14. Does the subject have acquired, clinically important alterations in behavior, personality, or compoment consistent with bvFTD of a neurodegenerative type? If answer is “No”, check “0 (Absent)” for Questions 15–21 and “0 (Meets <3 of the features described in Questions 15–21: does not meet criteria for bvFTD; or an exclusionary feature is present.)” for Question 22.	<input type="checkbox"/> 0	<input type="checkbox"/> 1

Characterizing symptoms of bvFTD				
<i>Have the following symptoms/behaviors been prominent, persistent, and recurrent in (approximately) the past three years?</i>	Absent	Questionably present	Definitely present	Not evaluated
15. Disinhibition 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"/> 9
16. Apathy or inertia 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"/> 9
17. Loss of sympathy/empathy Diminished response 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"/> 9
18. Ritualistic / compulsive behavior Simple repetitive movements or complex compulsive or ritualistic behaviors	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
19. Hyperorality and appetite changes 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"/> 9
20. Changes on neuropsychological testing consistent with bvFTD (refer to neuropsychological evaluation and neuropsychologist’s impression)	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9
21. Impaired daily functioning Are these alterations in behavior, personality, or compoment the principal cause of impaired daily living activities?	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 9

<p>22. Does the subject meet the criteria for clinical probable* or possible** bvFTD syndrome?</p> <p>*PROBABLE: Meets three of the above criteria and has impaired daily functioning and has imaging consistent with bvFTD.</p> <p>**POSSIBLE: Meets three of the above criteria but is not functionally impaired or does not having imaging consistent with bvFTD.</p> <p><i>NOTE: The diagnostic criteria in this module do not match the criteria in UDS V2.0 (Form D1). While Version 2.0 of the UDS is still in use, keep the two sets of diagnostic criteria separate.</i></p>	<p><input type="checkbox"/> 0 Meets <3 of the features described in Questions 15–21: does not meet criteria for bvFTD; or an exclusionary feature is present.</p> <p><input type="checkbox"/> 1 Probable bvFTD.</p> <p><input type="checkbox"/> 2 Meets criteria for possible bvFTD and has impaired daily functioning but without evidence of diagnostic imaging.</p> <p><input type="checkbox"/> 3 Meets criteria for possible bvFTD (with or without evidence of diagnostic imaging), but daily functioning is not significantly impaired.</p>
--	--

	No	Yes	Uncertain
23. Was an electromyogram (EMG) performed at this visit? If answer is "1 (Yes)", SKIP TO QUESTION 25.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	
24. Has an EMG been performed in the past year? If answer is "0 (No)", SKIP TO QUESTION 26.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	
25. If an EMG was performed, did it show evidence of motor neuron disease?	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 9

<p><i>If subject has only one diagnosis (either PPA or bvFTD), then END FORM HERE.</i></p>	
<p>26. For subjects with a diagnosis of both PPA and bvFTD, which diagnosis appeared first?</p>	<p><input type="checkbox"/> 1 bvFTD</p> <p><input type="checkbox"/> 2 PPA, semantic variant</p> <p><input type="checkbox"/> 3 PPA, nonfluent/agrammatic variant</p> <p><input type="checkbox"/> 4 PPA, logopenic variant</p> <p><input type="checkbox"/> 5 PPA not otherwise specified</p> <p><input type="checkbox"/> 9 Unknown</p>