

Criteria for Behavioral Variant FTD

In 2011, an international consortium developed revised guidelines for the diagnosis of behavioral variant frontotemporal dementia based on recent literature and collective experience. The following chart delineates the new criteria for bvFTD.

International consensus criteria for behavioural variant FTD

I. Neurodegenerative disease

The following symptom must be present to meet criteria for bvFTD

- A. Shows progressive deterioration of behaviour and/or cognition by observation or history (as provided by a knowledgeable informant).

II. Possible bvFTD

Three of the following behavioural/cognitive symptoms (A–F) must be present to meet criteria. Ascertainment requires that symptoms be persistent or recurrent, rather than single or rare events.

- A. Early* behavioural disinhibition [one of the following symptoms (A.1–A.3) must be present]:

- A.1. Socially inappropriate behaviour
- A.2. Loss of manners or decorum
- A.3. Impulsive, rash or careless actions

- B. Early apathy or inertia [one of the following symptoms (B.1–B.2) must be present]:

- B.1. Apathy
- B.2. Inertia

- C. Early loss of sympathy or empathy [one of the following symptoms (C.1–C.2) must be present]:

- C.1. Diminished response to other people's needs and feelings
- C.2. Diminished social interest, interrelatedness or personal warmth

- D. Early perseverative, stereotyped or compulsive/ritualistic behaviour [one of the following symptoms (D.1–D.3) must be present]:

- D.1. Simple repetitive movements
- D.2. Complex, compulsive or ritualistic behaviours
- D.3. Stereotypy of speech

- E. Hyperorality and dietary changes [one of the following symptoms (E.1–E.3) must be present]:

- E.1. Altered food preferences
- E.2. Binge eating, increased consumption of alcohol or cigarettes
- E.3. Oral exploration or consumption of inedible objects

- F. Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions [all of the following symptoms (F.1–F.3) must be present]:

- F.1. Deficits in executive tasks
- F.2. Relative sparing of episodic memory
- F.3. Relative sparing of visuospatial skills

III. Probable bvFTD

All of the following symptoms (A–C) must be present to meet criteria.

- A. Meets criteria for possible bvFTD
- B. Exhibits significant functional decline (by caregiver report or as evidenced by Clinical Dementia Rating Scale or Functional Activities Questionnaire scores)
- C. Imaging results consistent with bvFTD [one of the following (C.1–C.2) must be present]:
 - C.1. Frontal and/or anterior temporal atrophy on MRI or CT
 - C.2. Frontal and/or anterior temporal hypoperfusion or hypometabolism on PET or SPECT

IV. Behavioural variant FTD with definite FTLD Pathology

Criterion A and either criterion B or C must be present to meet criteria.

- A. Meets criteria for possible or probable bvFTD
- B. Histopathological evidence of FTLD on biopsy or at post-mortem
- C. Presence of a known pathogenic mutation

V. Exclusionary criteria for bvFTD

Criteria A and B must be answered negatively for any bvFTD diagnosis. Criterion C can be positive for possible bvFTD but must be negative for probable bvFTD.

- A. Pattern of deficits is better accounted for by other non-degenerative nervous system or medical disorders
- B. Behavioural disturbance is better accounted for by a psychiatric diagnosis
- C. Biomarkers strongly indicative of Alzheimer's disease or other neurodegenerative process

*As a general guideline 'early' refers to symptom presentation within the first 3 years. Table reprinted from *Brain*; permission granted to AFTD from Copyright Clearance Center. Rascofsky K et al. *Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia*. *Brain*. 2011 Sep, 134(pt9): 2456-77. Epub 2011 Aug 2.