

# Towards uniformity in bvFTD Phenocopy Syndrome: specialists' perspectives and proposed research criteria

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## BACKGROUND

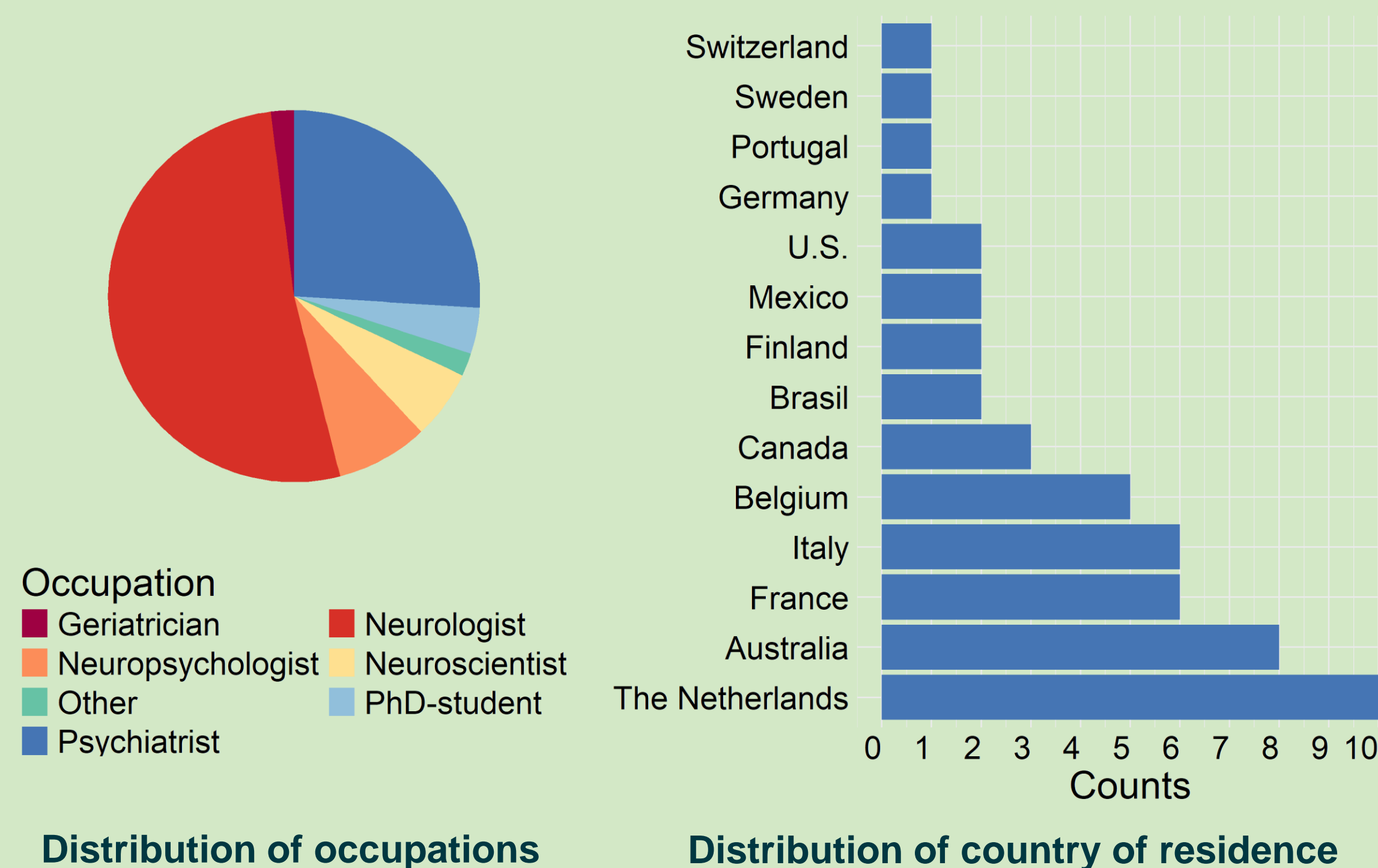
- The phenocopy syndrome of behavioral variant FTD (phFTD) refers to patients exhibiting clinical characteristics of bvFTD but without objective progression during follow-up
- Currently, diagnostic guidelines are lacking

## AIM

- To assess clinicians' perspectives on the diagnostic process for phFTD
- To develop research criteria for phFTD, by integrating these perspectives

## METHODS

- Establishment of the Phenocopy Working Group within the Neuropsychiatric International Consortium on Frontotemporal Dementia (NIC-FTD)
- International Delphi methodology:
  - 3 rounds: online survey + group discussion
  - 50 respondents



## PRELIMINARY RESEARCH CRITERIA

### Inclusion criteria

#### Baseline:

- Shows **deterioration of behavior and / or cognition** by observation or history as provided by a knowledgeable informant
- Age of onset of behavioral changes **> 35 years**
- At least **2 out of 6 core behavioral features of bvFTD** criteria (Rascovsky et al. 2011), either persistent or recurrent, rather than single or rare events.
  - Behavioral disinhibition
  - Apathy or inertia
  - Loss of empathy or sympathy
  - Perseverative, stereotyped, or compulsive or ritualistic behavior
  - Hyperorality and dietary changes
  - Neuropsychological profile: executive deficits with relative sparing of memory and visuospatial functions
- The **structural MRI is normal** or shows no abnormalities that could be explanatory for the behavioral change
- If available:
  - Absence of C9orf72** repeat expansion carriership
  - Normal NfL or AD biomarker levels** in CSF or plasma (unless elevated levels can be explained by another factor known to elevate levels)
  - FDG-PET not suggestive of neurodegeneration**

#### Follow-up:

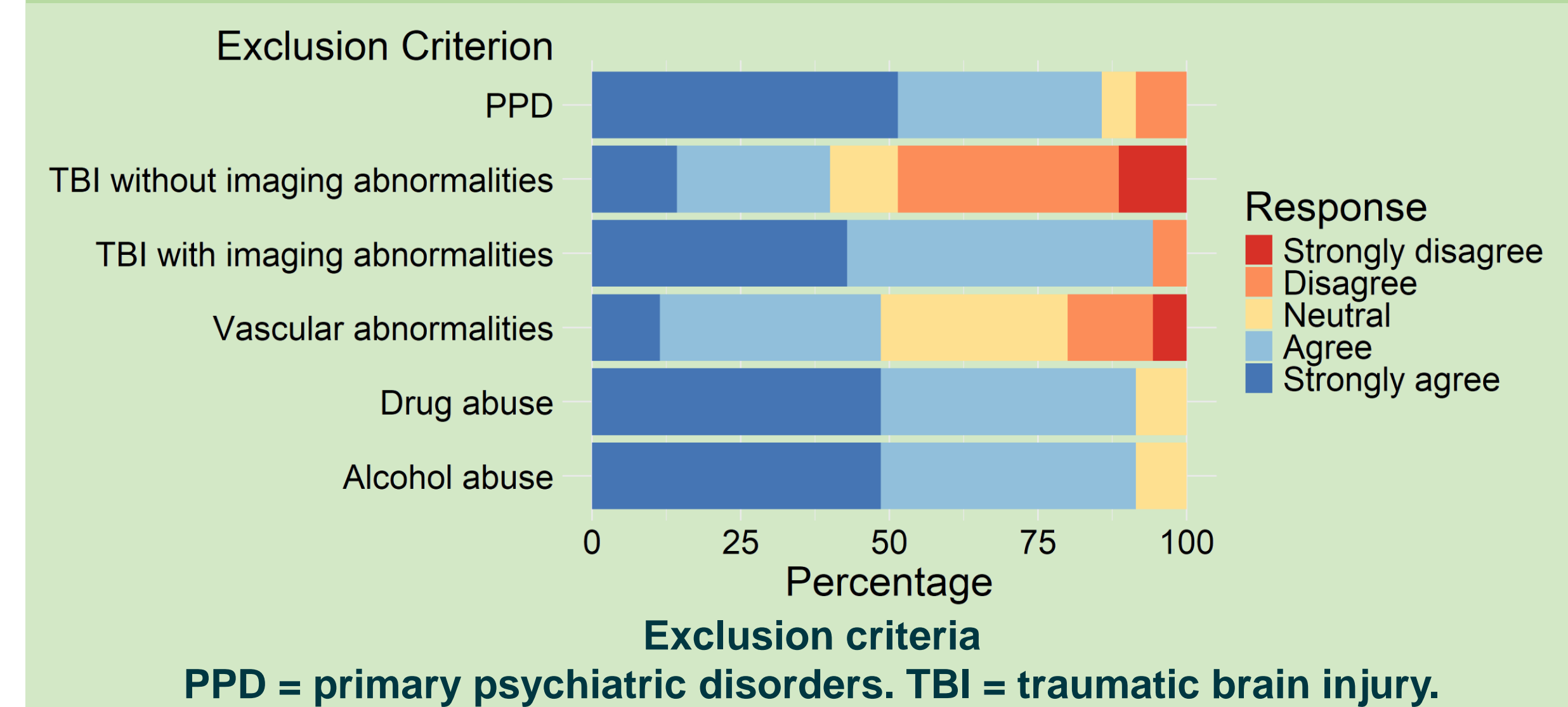
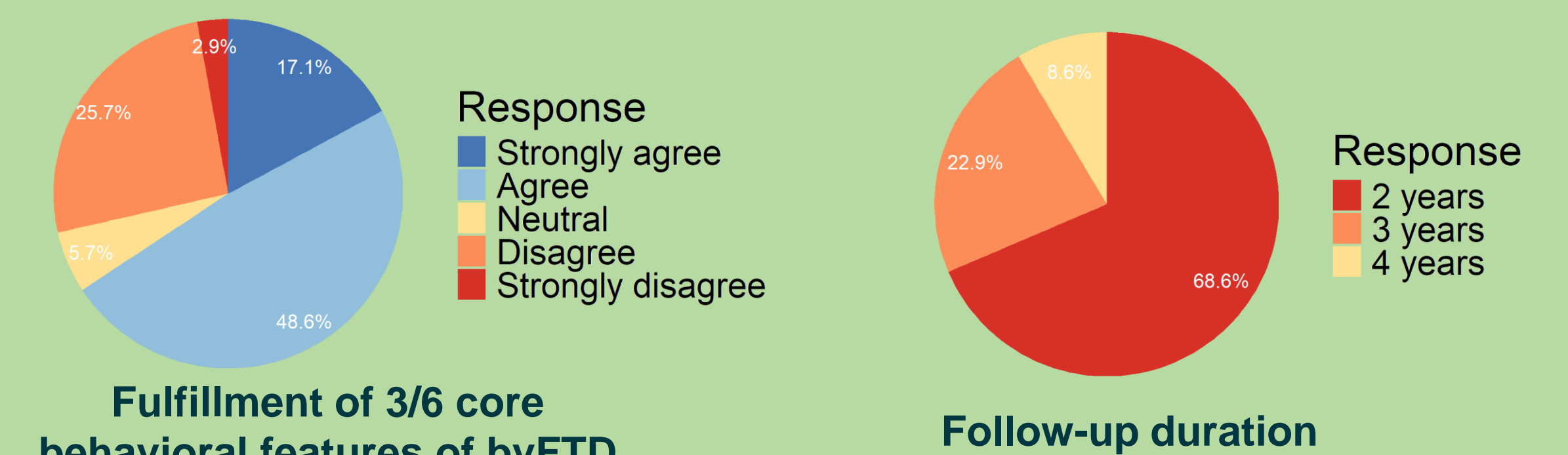
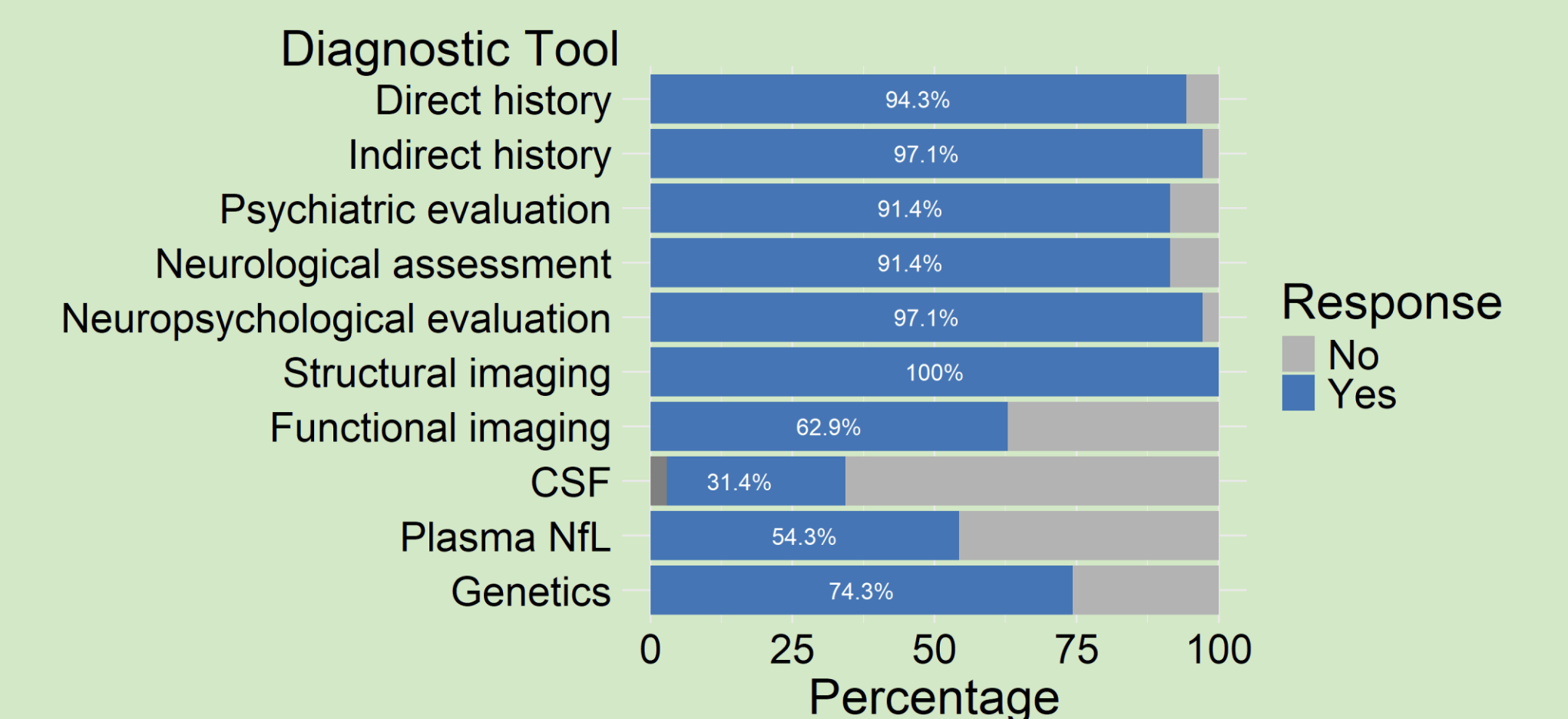
#### Absence of objective progression over at least 2 years:

- Absence of progression on **neuropsychological evaluation**; AND
- Absence of progressive abnormalities on **structural MRI** (or FDG-PET)

### Exclusion criteria

- The symptoms are better accounted for by **another neurological or psychiatric disorder** (according to the DSM-5), including alcohol or drug abuse
- Presence of **traumatic brain injury** that better accounts for the behavioral disturbance
- Fulfilment of radiological criteria for **vascular cognitive impairment**

## SURVEY RESULTS



## CONCLUSION

This study will result in a set of phFTD research criteria

## NEXT STEPS

International prospective cohort study

- Prospective data collection on phFTD cases
- Identify contributing factors & characteristic features
- Improved insight into the etiology