A cross-sectional and longitudinal investigation of the Cambridge Behavioural Inventory – Revised (CBI-R) in behavioural variant frontotemporal dementia (bvFTD)

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BACKGROUND
Changes in behavioural and neuropsychiatric profiles are hallmark features of behavioural variant frontotemporal dementia (bvFTD). Informant based questionnaires that reliably capture these deficits in bvFTD, such as the Cambridge Behavioural Inventory-Revised (CBI-R), are commonly used in clinical research and increasingly considered for therapeutic trials.

While studies have investigated the cross-sectional validity of the CBI-R in bvFTD, little is known about how bvFTD compares with the primary progressive aphasia (PPA) variants of FTD. In addition, the longitudinal feasibility of these scales is currently lacking.

The current study therefore investigated the cross-sectional CBI-R profiles in the three canonical variants (bvFTD, semantic variant PPA (svPPA) and non-fluent PPA (nfvPPA)) and assessed its utility as a longitudinal biomarker of disease progression.

METHODS
The CBI-R was completed by primary caregivers for 63 FTD patients (Table 1). For each of the 10 FTD domains (see Figure 2) scores were converted to percentage of maximum score. Total CBI-R is scored out of 180 with higher numbers indicating greater deficits. 29 FTD patients returned for longitudinal investigations (Table 1).

RESULTS: CROSS-SECTIONAL ANALYSIS
Across all 10 CBI-R domains bvFTD demonstrated the greatest deficits (Figure 1). The most severely affected domains were 'Stereotypic and Motor Behaviours', 'Memory and Orientation', 'Motivation' and 'Eating Habits' at 58.0%, 57.5%, 55.6% and 49.5% of maximum possible score respectively. The least affected domains were 'Beliefs' and 'Self Care' at 7.3% and 15.6%. The svPPA group also showed significant deficits in multiple domains. Although scores were lower than the bvFTD group, they were not significantly different. nfvPPA demonstrated fewer deficits with significantly lower scores in multiple domains (Table 2).

RESULTS: LONGITUDINAL ANALYSIS
Mean (sd) annual percentage change in CBI-R scores are shown in Table 3. The domains which exhibited the greatest annualised change were 'Self Care' for bvFTD, 'Motivation', 'Memory and Orientation', 'Everyday Skills' and 'Eating Habits' for svPPA and 'Stereotypic and Motor Behaviours' for nfvPPA, although the large variability in each domain resulting in a non-significant difference (Figure 2). Total CBI-R score was also variable, demonstrating an overall increase in only 9/18 bvFTD, 6/7 svPPA and 3/4 nfvPPA patients (Figure 3).

CONCLUSIONS
CBI-R proved useful as a cross-sectional marker in multiple symptomatic domains across the FTD spectrum. bvFTD demonstrated the greatest deficits across all domains, although there were also significant deficits within the svPPA group. The longitudinal variability of CBI-R scores underlines it as a marker of disease progression. This highlights the importance of validating biomarkers longitudinally for clinical research and prior to inclusion as outcomes in putative therapeutic trials.