The Cambridge Behavioural Inventory (Revised) detects early behavioural and functional impairment in genetic frontotemporal dementia within the GENFI cohort.

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Background

Behavioural dysfunction is a key feature of frontotemporal dementia (FTD) but validated clinical scales measuring behaviour are lacking at present. The Cambridge Behavioural Inventory – Revised (CBI-R) is a 41-item questionnaire measuring the severity of behavioural and functional issues commonly seen in FTD that is self-administered, consisting of 10 domains (Memory and Orientation, Everyday Skills, Self Care, Mood, Abnormal Behaviour, Eating Habits, Sleep, Beliefs, Stereotypic Behaviour and Motivation). The aim of this study was to assess the CBI-R questionnaire as a measure of disease severity in genetic FTD.

Methods

We assessed behaviour using the revised version of the Cambridge Behavioural Inventory (CBI-R) in 733 participants from the Genetic FTD Initiative study: 466 mutation carriers (195 C9orf72, 76 MAPT, 195 GRN) and 267 non-mutation carriers (healthy controls). All mutation carriers were stratified according to their CDR® plus NACC FTLD into three groups: asymptomatic (CDR=0), mildly symptomatic (CDR=0.5) and fully symptomatic (CDR=1+). CBI-R total scores were compared between mutation carrier groups using a mixed effects model that adjusted for age, education, sex and family clustering, with 95% bootstrapped confidence intervals with 2000 repetitions to adjust for non-normally distributed data. We used the same mixed effects model to run a within-group analysis between the 10 CBI-R domains in CDR 1+ groups. Spearman rank correlations were run to assess the relationship of the CDR® plus NACC FTLD SOB and FRS scale with the CBI-R total scores in CDR 1+ mutation carrier groups.

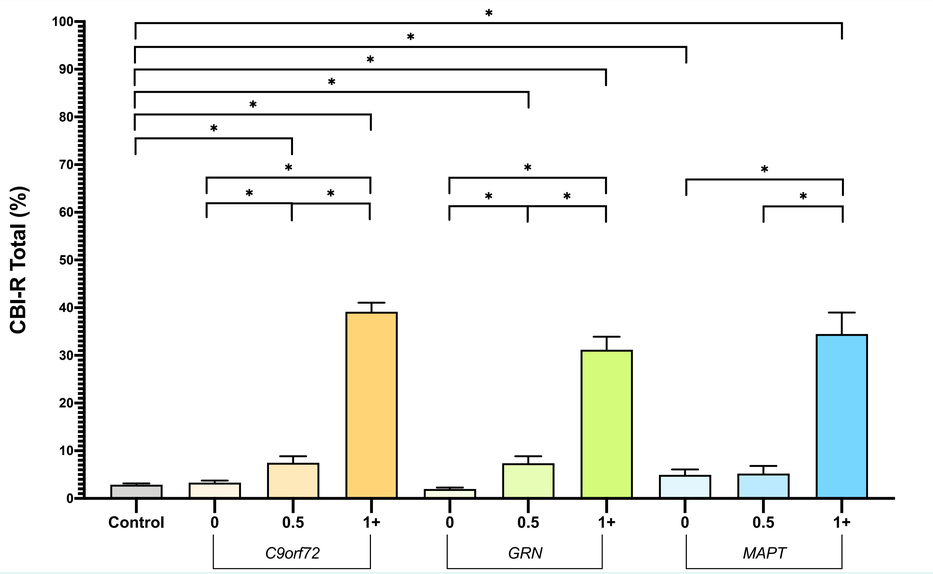

Figure 1.

Results

CBI-R total scores were significantly higher in all CDR 1+ mutation carrier groups compared to controls (C9orf72 mean 70.5 (standard deviation 27.8), GRN 56.2 (33.5), MAPT 62.1 (36.9)), as well as their respective CDR 0.5 groups (C9orf72 13.5 (14.4), GRN 13.3 (13.5), MAPT 9.4 (10.4)) and CDR 0 groups (C9orf72 6.0 (7.9), GRN 3.6 (6.0), MAPT 8.5 (13.3)). Both C9orf72 and GRN 0.5 groups scored significantly higher than controls and their respective CDR 0 groups **(see Figure 1).**

Motivation and Memory were the highest scoring domains in both C9orf72 and GRN CDR 1+ groups, and in the MAPT group the highest scoring domains were Stereotypic Behaviour and Memory.

There was a positive correlation between the CBI-R total scores and the CDR ® plus NACC FTLD SOB scores in all mutation carrier groups (C9orf72: rho= 0.8, p<0.001, GRN: rho= 0.8, p<0.001, MAPT: rho= 0.6, p<0.001), and a negative correlation between CBI-R total scores and FTD Rating Scale scores (C9orf72; rho= -0.9, p<0.001, GRN; rho= -0.9, p<0.001, MAPT; rho= -0.9, p<0.001) **(see Figure 2)**.

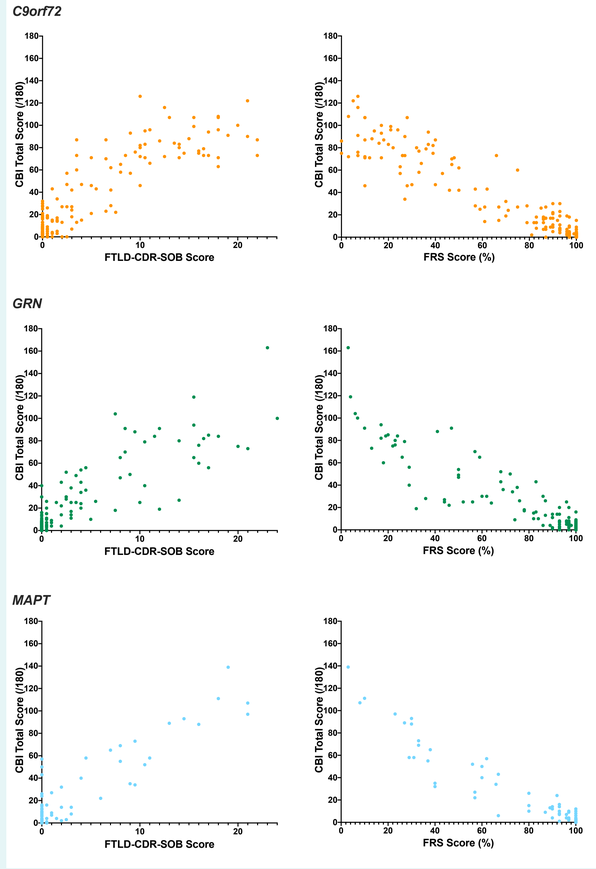

Figure 2.

Conclusions

The CBI-R detects early behavioural change in genetic FTD, particularly in those with C9orf72 and GRN mutations. The CBI-R could be a useful marker within a clinical trial setting.

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