Preclinical signs of impairment in persons at high risk of frontotemporal dementia related to chromosome 3 (FTD3): Preliminary findings in neuropsychological tests

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BACKGROUND
We have studied a large kindred in Jutland with autosomal dominant frontotemporal dementia linked to chromosome 3, the only such family yet known. 1985-7: Initial studies (Gydesen et al., 1987) 1995: Linkage to the pericentromeric region of chromosome 3 (Brown et al., 1995) 2002: Publication of full clinical details (Gydesen et al., 2002)

METHODS
At risk members of the family and their spouses between 40 and 70 years of age were invited to participate in neuropsychological assessment performed without knowledge of status. 38 family members and 20 spouses participated. Some of the subjects have not yet been haplotyped, and we report preliminary results from comparisons of 20 test measures in 3 well-matched subject groups: 11 high risk subjects, 16 low risk subjects, and 19 spouses.

RESULTS
T-tests without corrections for multiple comparisons showed:
1) No significant differences between the two control groups;
2) A total of 8 significant differences (2 at the .002 level) between high risk family members and controls, all with the high risk subjects impaired.

Trail Making B was impaired relative to both control groups, and significant differences between high risk subjects and one control group (but not both) were found in cognitive estimations, letter-number sequences (a measure of working memory control), immediate (but not delayed) story recall, and one further test (table 1).

CONCLUSION
The pattern of subtle impairments found in the high risk family members is compatible with predominantly frontal involvement. These preliminary results must be confirmed in the full data set and replicated in further cross-sectional and longitudinal analyses in a follow-up, which has been planned for this Autumn.

REFERENCES

OBJECTIVE
The phenotype in the early phase of the disease is not yet known, and we studied well subjects at risk of developing the disease to detect early signs

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