

Gender Differences in the Clinical Presentation of Familial Frontotemporal Dementia

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INTRODUCTION

A large kindred was identified with a high prevalence of frontotemporal dementia (FTD), a neurodegenerative disorder that causes primarily behavioral, language and social dysfunction. The age of onset for this disease ranges from the 40s to the 60s. The disorder appears to have a frontal onset followed by frontal and temporal lobe dysfunction. Parkinsonism and motor neuron disease have not been observed.

SUBJECTS

- Large kindred with high prevalence of FTD
- A majority of subjects were right handed (100%), Caucasian (100%) and female (56%)

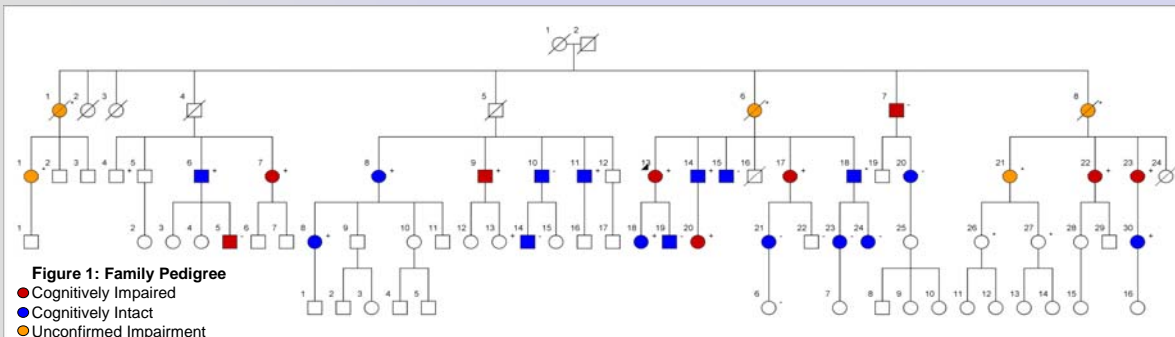
	Range	Mean	SD
Age at Test	24 - 86	48.4	14.5
Education	8 - 19	12.7	2.7

Table 1
Demographics of Cohort (N=25)

METHODS

- Comprehensive neuropsychological testing included:
 - Basic Orientation: Mini-Mental State Exam
 - Pre-Morbid Intelligence Estimates: Wide Range Achievement Test-Revised, WAIS III subtests: Comprehension (Comp), Information (Info)
 - Executive Function: Rey-O Complex Figure, Hooper, Stroop Color-Word, Wisconsin Card Sort, Trails B, Matrix Reasoning (MR), Similarities, Fluency (FAS), Picture Completion, Picture Arrangement (PA)
 - Attention: Digit Span (Dspan), Verbal Series Attention Test (VSAT)
 - Information Processing: Trails A, Symbol-Digit Modality (SDM)
 - Visuospatial Ability: Benton Faces, Judgment of Line Orientation (JLO), Block Design (BlockD),
 - Semantic Fluency: Animals
 - Episodic Memory: Rey Auditory-Verbal Learning Test, Brief Visuospatial Memory Test
 - Naming: Boston Naming Test
 - Fine Motor: Grooved Pegboard
 - Depression: Beck Depression Inventory (BDI-II)
- Statistical analysis to determine cognitive impairment

GENEALOGY OF FAMILY



RESULTS

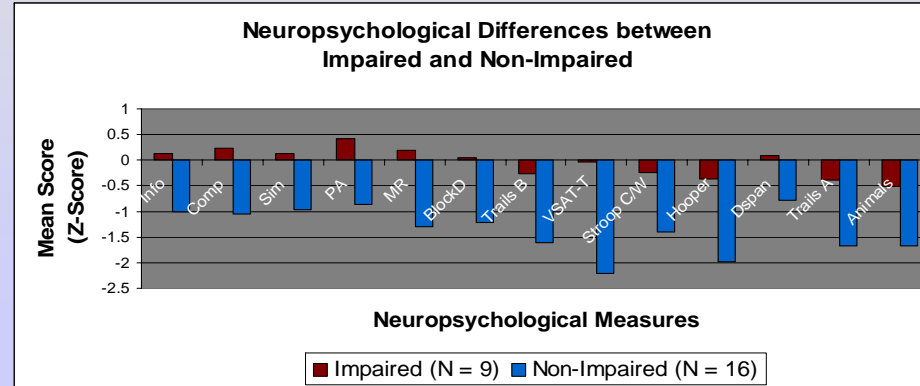


Figure 2 Differences between Impaired and Non-Impaired

The mean z-scores for both groups was found. The above tests, which measure executive function, attention, working and semantic memory, and visuospatial skills were found to be significant different between groups ($p < 0.05$ in all cases). Frequency of gender and depression (BDI) were not different between Impaired and Non-Impaired groups.

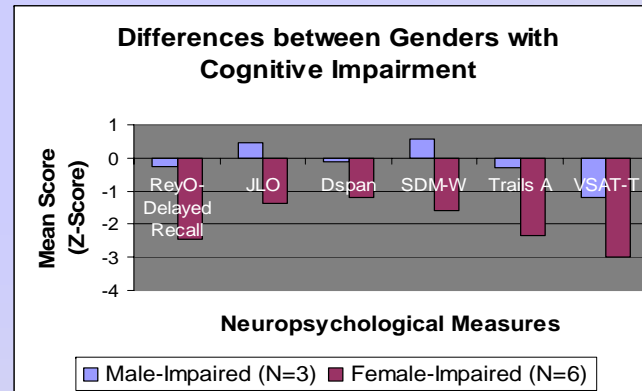


Figure 3 Differences between Cognitive Impaired Males and Females

The mean z-scores for cognitively impaired males and females was found across a variety of cognitive domains. Females performed significantly worse than males ($p < 0.05$ in all cases). Depression (BDI) was not different between genders.

CONCLUSIONS

Comprehensive neuropsychological testing of affected persons has revealed problems in the cognitive domains of executive function, attention, working and semantic memory, and visuospatial skills. Within cognitively impaired family members, women have worse cognition than men. Medial temporal functions are unaffected as would be expected in FTD. The family appears to be unique in having different clinical presentations based on gender.

The cognitive impairment found in our familial FTD cohort is consistent with a typical FTD pattern, highlighting impairments in attention and executive functioning with intact memory abilities. Although results are preliminary given the small sample size, findings present a unique clinical phenotype in terms of the severity of cognitive impairments found in the female members as compared to their male counterparts. Future work will involve characterizing additional family members to ascertain this family's clinical phenotype and to identify the genetic mutation underlying their disorder.