Original Article

Personality Traits in the Siblings and Children of Patients with Frontotemporal Dementia: A Questionnaire-based Study

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ABSTRACT

Introduction: Frontotemporal dementias (FLD) form a group of relatively young onset, male dominant dementias with significant behavioral abnormalities early in the course of the disease. Routine assessment suggested preexisting traits such as lack of empathy, self-directedness, and persistence in most of these persons even before the onset of disease. Hence, we decided the study, the siblings and children of patients for any specific traits and correlation with hexanucleotide expansion repeats if any traits were identified. **Patients and Methods:** A total of 35 age- and gender-matched cases and controls were included for the study as per criteria. They were screened for mental illness and cognitive dysfunction using Hindi Mental State Examination and Mini-mental State Examination. Eligible persons were given temperament and character inventory (TCI) scores for the recommended parameters. Hexanucleotide expansion was also studied in the patients, cases and controls. **Results:** No specific personality trait was found to have an increased correlation with siblings and children of patients with FLD in this small group using TCI scores. **Conclusions:** 7% of cases showed Hexanucleotide expansion suggesting a possible risk.The role of self reporting bias resulting in normal personality trait needs to be addressed in future studies.

Key words: *Frontotemporal dementia, personality traits in close family members, temperament and character inventory scores*

INTRODUCTION

Frontotemporal dementia (FLD) is the term applied to a heterogeneous group of cortical dementias, which is seen in relatively young males more than females.

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Mostly, it is a clinical syndrome of behavioral changes with memory impairment with or without early language change depending on the mode of onset of disease. There are three main types: The behavioral variant, progressive aphasia type, and the semantic variant. The visuospatial functions, episodic memory, and orientation are well-preserved; therefore, they fare relatively well in laboratory tests but fail in life due to intractable behavioral changes. In a study conducted in our department, we found that only 11% of our patients had memory changes in the early stage as against 25% in the literature. Therefore, a diagnosis of FLD is delayed in our patients (our study ahead of print) social, and personal skills and behavior are lost early with disinhibition, mental inflexibility but with maintained independence. There is a significant deterioration in interpersonal conduct, emotional modulation, the presence of stereotypical and aberrant motor behavior, changes in eating preferences which indicate progressive deterioration in neural circuits concerned with social cognition. The beginning is very difficult to understand as it is more behavioral and not intellectual.

Early diagnosis in patients is assisted by the development of carer-based questionnaires designed to document the range of symptoms found in the person with dementia, including the neuropsychiatric inventory, Cambridge behavioral inventory, and frontal behavioral inventory.[1] Routine assessment of our patients revealed that most of our patients had distinct personality traits before gross social deterioration took place. They appeared to be disciplinarians who lacked empathy, self-directed (SD), cold, and obstinate. Therefore, we decided to conduct a questionnaire-based self-rated study of personality traits in children and siblings of diagnosed patients with behavioral variant FLD. Nearly 40% of patients have a positive family history,[2] and therefore, a very high possibility of detecting certain traits was hypothesized assesses using temperament character inventory (TCI).[3] This proposes four temperament dimensions (1) novelty seeking (NS), (2) harm avoidance (HA), (3) reward dependent (RD), (4) persistence (PS) and three characters. (1) SD (2) self-transcendence (ST) (3) cooperativeness (CO) these traits are believed to be stable during life and is based on the psychobiological model that attempts to explain the underlying causes of individual differences in personality traits. Low basal dopamine correlates with decreased NS, which correlates with extroversion, openness to experience, and impulsivity, HA with increased serotonin which correlates with neuroticism, decreased noradrenalin with RD. PS is associated with conscientiousness, CO with agreeableness, and SD has a negative association with neuroticism. Cloning argues that psychological well-being correlates with autonomy, life purpose, and positive relationships.

Soderstrom *et al*.[4] reported an association between the increased ratio of homovanillic acid serotonin metabolites with traits who lack empathy, guilt, and remorse which might be due to disinhibition of violent antisocial traits due to impaired serotonin regulation of dopamine. There is also role via hypothalamo-pituitary-adrenal axis resulting in serotonin based dysregulation causing low cortisol to stress, increase the levels of testosterones resulting in dominance seeking and aggression. Cortisol prepares the system to provide energy during stress, potentiates fear, sensitivity to punishment, and withdrawal.[5] Reduced volume of the amygdala is reported in patients who lack learning of the sufferings caused to others by their behavior. This probably occurs early prenatal period or childhood due to the influence of hormonal imbalance.[6] Hippocampus which has dense links to the amygdala and prefrontal cortex also shows abnormalities.[7] Other structures and connectivity postulated are anterior cingulum, parahippocampal region, anterior superior temporal gyrus, insula, etc.[8] Decision-making and cognitive evaluation may be affected by disruption of connections between subcortical structures to the cortex.[9] Therefore, it is likely that subtle personality changes might serve as markers for underlying neurochemical and connectivity associated changes which might predispose the person later to neurodegenerative processes was our hypothesis.

PATIENTS AND METHODS

The aims and objectives were to know whether any particular personality trait is seen in apparently normal family members of patients with FTD. An attempt will be made to find out association with known gene mutation like hexanucleotide repeat expansion GGGGCC. If any specific trait is identified, it might serve as a biomarker of early diagnosis. Early diagnosis might facilitate interventions to postpone the onset of symptoms.

Siblings and children of probable patients with behavior variant FLD as per international consensus criteria, who were willing to participate in the study were included. Their age must be above 18 years. Their cognitive status should be normal as per Hindi mental status examination and not suffering from mental illness as per miniscreen for mental illness.[10] This consists of three sections. Section A for mood disorders, Section B for anxiety and Section C for psychotic disorders it has 22 items which need yes or no answers. All positive answers are added up and total score and score in each section is taken if the patient has suicidality irrespective of score needs evaluation. If question 14 and 15 are yes, it means posttraumatic stress disorder. If the total score is <5 low likelihood of mental illness. Those who had a score of

6–9 are having a moderate risk and more than 9 high risk. Only those who qualified for low risk were included in the study. Children <18 years and unwilling persons were excluded from the study. They were evaluated with TCI-revised personality questionnaire. This consists of the following parameters. NS, HA, RD, PS [Tables 1-4], self-directiveness, ST, CO, 10 ml of venous blood was drawn to study the prevalence of hexanucleotide expansion in the study group and the control group. Consist of GGGGCC repeats in the noncoding region of chromosome 9p21 it is inherited in a dominant pattern. It is commonly associated with FTD amyotrophic lateral sclerosis. It generally causes disease by toxic gain of function due to the accumulation of noncoding segment of RNA. Infrastructure was not available for the study of other genotypes. Control consisted of persons who satisfied all the above criteria but did have a parent or sibling with behavioral variant FTD.

Ethical clearance was obtained from the Institutional Ethical Committee for the above study. There were 35 persons in the study group and 35 persons in the control group. DNA isolation was done as per Millers *et al*. method 1988 the study group consisted of siblings and children of patients and control group consisted of age- and gender-matched bystanders of patients with other noncognitive disorders. The revised temperament character scale was applied to all of them. The information obtained was compared statistically and analyzed using *t*-test, Chi-square test.

Table 1: Demographic data

	Patient Relative	Control
Age	32.77±6.76	32.77±6.76
Gender	11/24	11/24
Ethnicity (north/south)	6/29	6/29

Table 2: Novelty Seeking (NS) parameter of temperament character inventory between cases and controls

	Patient Relative	Control	*P*-Value
NS1	24.57±3.91	24.86±3.98	0.88
NS2	20.46±7.83	20±4.26	0.99
NS3	19.17±3.09	19.17±2.87	0.98
NS4	18.6±3.6	18.4±3.7	0.7
Total	82±11.29	82±7.7	0.91

Table 3: Harm avoidance parameter of temperament character inventory between cases and controls

	Patient relative	Control	*P*-value
HA1	23.86±5.78	24.51±4.55	0.74
HA2	18.54±3.4	19.46±2.3	0.25
HA3	14.4±4.9	16.31±2.4	0.20
HA4	17.6±4.9	18.26±3.4	0.95
Total	74.26±15.8	78.89±16.7	0.72

Table 4: Cooperativeness parameter of temperament Character Inventory (CI) between cases and controls

	Patient relative	Control	*P*-value
C1	22.37±5.1	22.03±5.1	0.81
C2	16.03±1.82	16.17±2.21	0.94
C3	23.51±4.54	22.71±3.65	0.81
C4	19.91±4.55	20.06±4.64	0.93
C5	20.94±3.79	21.77±3.24	0.32
Total	102.86±13.49	102.71±11.23	0.97

International consensus criteria for behavioral variant of frontotemporal dementia

Neurodegenerative disease

It must be present for any FTD clinical syndrome and shows progressive deterioration of behavior and/or cognition by observation or history.

Possible behavioral variant of frontotemporal dementia

Three of the features (a-f) must be present; symptoms should occur repeatedly, not just as a single instance:

a. Early (3 years) behavioral disinhibition
b. Early (3 years) apathy or inertia
c. Early (3 years) loss of sympathy or empathy
d. Early (3 years) perseverative, stereotyped, or compulsive/ritualistic behavior
e. Hyperorality and dietary changes
f. Neuropsychological profile: Executive function deficits with relative sparing of memory and visuospatial functions.

Probable behavioral variant of frontotemporal dementia

All the following criteria must be present to meet diagnosis:

a. Meets criteria for possible behavioral variant of FTD (bvFTD)
b. Significant functional decline
c. Imaging results consistent with bvFTD (frontal and/or anterior temporal atrophy on computed tomography (or) magnetic resonance imaging (or) frontal hypoperfusion (or) hypometabolism on single-photon emission computed tomography or positron emission tomography).

Definite behavioral variant of frontotemporal dementia

Criteria a, either b or c must be present to meet diagnosis:

a. Meets criteria for possible or probable bvFTD
b. Histopathological evidence of FTLD on biopsy at postmortem
c. The presence of a known pathogenic mutation.

Exclusion criteria for behavioral variant of frontotemporal dementia

Criteria a and b must both be answered negatively, Criteria c can be positive for possible bvFTD but must be negative for probable bvFTD:

a. Pattern of deficits is better accounted for by other

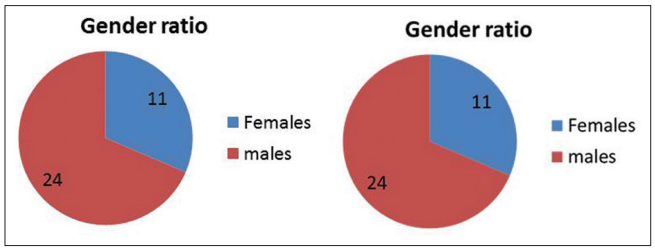

Figure 1: Indicates male female ratio between cases and controls

nondegenerative nervous system (or) medical disorders
b. Behavioral disturbance is better accounted for by a psychiatric diagnosis
c. Biomarkers strongly indicative of Alzheimer's disease or other neurodegenerative process.

Additional features

a. Presence of motor neuron findings suggestive of motor neuron disease
b. Motor symptoms and signs similar to corticobasal degeneration and progressive supranuclear palsy
c. Impaired word and object knowledge
d. Motor speech deficits
e. Substantial grammatical deficits.

RESULTS

There were 35 persons in both groups. Gender and ethnicity were exactly matched 11 females in each group and ethnicity too well-matched 6 north Indians in each group [Figures 1,2 and Table 1]. Mean age was 37.77 plus or minus 6 in both cases and controls. The TCI scores with reference to NS, HA, RD, PS, SD, ST, CO did not show any significant difference between cases and controls. With reference to hexanucleotide expansion 47 patients with FLD and their siblings were evaluated. Four patients showed more than thirty hexanucleotide expansion and one patient had more than 14 repeats. However, these persons family too did not show any significant personality trait as assessed using the above methods.

CONCLUSION

This study using self-rated questionnaire TCI does not reveal and specific personality traits in close relatives of patients with FLD, which can serve as a predictive biomarker for future occurrence of FLD. Hexanucleotide expansion more than 30 repeats was seen in about 7.5% of patients with FLD in this group analyzed and none in the control group; however, its prognostic role remains to be studied. There is self-reporting bias regarding personality traits, and the number is also small. Hence, a larger sample needs to be studied.

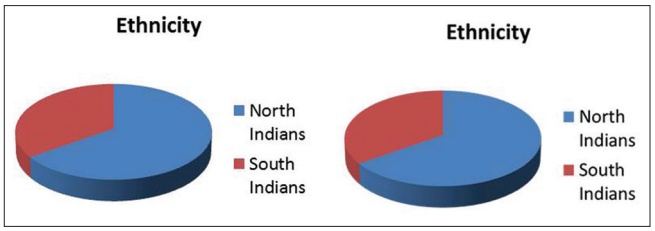

Figure 2: Indicates region to which the patient belongs

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Conflicts of interest

There are no conflicts of interest.

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