FRONTOTEMPORAL DEMENTIA: RECOGNIZING PATTERNS OF COGNITIVE PERFORMANCE IN RELATION TO ACTIVITIES OF DAILY LIVING

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Dedication

This thesis project is dedicated to the psychology department faculty at California State University Northridge, in particular, Drs. Jill Razani, Gary Katz and Mark Sergi. For without their wisdom, kindness, enthusiasm and encouragement this project would not be possible. A special thanks to Jessica Worland for her efforts in making this, and all endeavors, possible in the Psychology Department at CSUN.
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ABSTRACT

FRONTOTEMPORAL DEMENTIA: RECOGNIZING PATTERNS OF COGNITIVE PERFORMANCE IN RELATION TO ACTIVITIES OF DAILY LIVING

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Dementia represents a series of degenerative diseases known to cause a decline in cognitive functioning. A significant yet understudied type of dementia is frontotemporal dementia (FTD), characterized primarily by deficits in executive abilities and language. There is, particularly, little data regarding the daily functional abilities of FTD patients. The current study aimed to detail patterns of activities of daily living, using an observation-based test, and neuropsychological performance of patients with FTD. Thirteen participants with FTD were administered the Direct Assessment of Functional Status (DAFS). Fourteen subscales of the DAFS, designed to assess areas of orientation, communication, transportation, shopping, and financial skills were assessed. Each task within the subscales is observed and rated objectively by the researcher who scores participants on their performance; higher scores are equivalent to better performance. DAFS subscale scores were then converted to percentage correct responses for each subscale. FTD and 57 normal age- and education-matched controls also completed a
battery of cognitive tests designed to assess major cognitive domains, which included the Digit Span, CVLT, WCST, FAS and Rey-O. FTD patients’ raw neuropsychological test scores were converted to z-scores using control participants’ mean and standard deviation measures in order to create comparable units of measurement. The results revealed no significant differences between the neuropsychological z-scores for the FTD. However, a significant difference in ADL performance across groups, with NC outperforming those with FTD on nearly all areas of ADL performance was found. Within group analyses revealed that tasks related to recall, recognition and executive functioning were particularly difficult for those with FTD. Overall results support prior research in stating that those with FTD will be outperformed by normal controls on tasks related to ADL functioning. However, what this study specifically identifies is that within the range of deficits characterized through the DAFS scores, there are areas of ADL performance which are significantly more challenging for those with FTD in relation to not only age matched controls, but within their own patterns of deficits.


Introduction

Dementia: A General Overview

The term dementia is used to distinguish individuals with cognitive decline as a result of various disorders. As recognized by the National Institute of Neurological Disorders and Stroke (NINDS, 2012) dementia represents a collection of symptoms that ultimately, lead to an interruption in normal activities and relationships. Clinically, individuals with dementia may present difficulty with problem solving and emotional regulation, memory, and behavior modification. Other abilities that may be limited include language skills, perception and cognitive skills, including reasoning and judgment. According to NINDS (2012) at least two or more of these functions must be significantly impaired without the loss of consciousness, to be deemed as a form of dementia.

According to a study by Cleusa et. al (2006), approximately 24-30 million people worldwide are living with some form of dementia, with four to six million new cases each year. According to Cleusa (2006), this number is expected to double and even triple over the next 20 years, making research on this topic imperative. Listed as the 6th leading cause of death in the United States, Alzheimer’s Disease is the most common form of dementia known and is responsible for roughly 84,000 deaths each year (CDC, 2013). Further estimates by Plassman et. al (2007) suggest that roughly 13% of Americans over the age of 71 have some form of dementia, where 9.7% had Alzheimer’s Disease. These findings further showed that roughly 40% of those over age 90 are likely to develop Alzheimer's disease if not a closely related dementia (Plassman et. al, 2007).
As further recognized by Plassman (2007), as individuals are living longer, dementia related diagnoses are also increasing. Overall, as previous studies demonstrate, there is a prevalent and distinct impact of dementia in and outside the U.S., where further research is needed. As such, the current study hopes to increase our understanding of the symptoms, patterns and outcomes associated with a particular form of dementia, Frontotemporal Dementia (FTD).

*Frontotemporal Dementia: Neuropathology, Cognitive Impairment and Symptom Patterns*

While much research has been devoted to the study of Alzheimer’s disease, there is a comparable yet understudied type of dementia, Frontotemporal Dementia (FTD). Although epidemiologic studies are limited, approximately 20 to 50 percent of persons younger than 65 years with dementia have FTD, a prevalence similar to that of Alzheimer disease in persons 45 to 64 years of age (15 per 100,000) (Cardarelli, 2010). Similar findings were found in a study by Ratnavalli et. al (2002), who evaluated 108 patients with potential FTD and AD. Overall, FTD can be distinguished from other forms of dementia through detailed history, physical examination, neuropsychological testing and the application and guidance of MRI scans (Cardarelli, 2010).

In general, FTD is the result of deterioration in the frontal and temporal lobes of the brain. As supported by Cardarelli (2010), neuroimaging (magnetic resonance imaging-MRI) usually demonstrates focal atrophy in these areas. In many cases, the atrophy (or cell degeneration) stems from an abnormal form of the TAU (Tubulin Associated Unit) protein in the brain, which leads to the accumulation of neurofibrillary tangles. The
development of tangles disrupts regular cell activity, which in turn, can cause cell
dysfunction and death (NINDS, 2012). The structures found in the frontal and temporal
lobes of the brain primarily control judgment and social behavior, therefore, as a result of
cell damage, people with FTD often have problems maintaining normal interactions and
following social conventions.

Individuals with FTD may display inappropriate behavior, socially odd behaviors, and
may neglect their normal responsibilities (NINDS, 2012). Other common symptoms
generalized to the FTD population include loss of speech and language, compulsive or
repetitive behavior, increased appetite and motor problems such as stiffness and balance
problems. Memory loss may also occur, although it typically appears later in the disease;
it is the preservation of memory that generally differentiates FTD from dementia of the
Alzheimer’s type along with its earlier age of onset (Cardarelli et. al, 2010).

Overall, as identified by the National Institute of Aging (NIA, 2013) the deficits found in
FTD can be grouped into three categories of impairment. The first is progressive
behavior/personality decline characterized by changes in personality, behavior, emotions
and judgment. The second is progressive language decline which is marked by early
changes in language ability, including speaking, understanding, reading and writing. The
final and third area of dysfunction includes progressive motor decline, characterized by
various difficulties with physical movement, including shaking, difficulty walking,
frequent falls and poor coordination (NIA, 2013).
In terms of neuropsychological performance, Braaten et al (2006) identified specific patterns of abilities across differing forms of dementia. FTD was distinguished from dementia of Alzheimer’s Type, Vascular Dementia, and also Major Depressive Disorder, which according to Braaten (2006), pose differential diagnostic challenges. Neuropsychological domains directly assessed by Braaten (2006) and his colleagues included immediate memory, delayed memory and confrontational naming. Other measures included verbal fluency, attention, concentration, and executive functioning. Overall results revealed distinct neuropsychological profiles for Dementia of Alzheimer’s Type, Vascular Dementia, Frontotemporal Dementia and Major Depressive Disorder where those with FTD in particular, performed significantly worse than the other groups on tests of executive functioning (Braaten et al, 2006).

Similar findings were seen in a study by Pachana et al (2006), where 15 FTD patients were compared to 16 AD patients and 16 controls. Overall results showed controls to outperform the AD and FTD groups on tests of verbal and nonverbal memory, executive ability and constructional skill, where AD patients held more widespread memory decline –nonverbal memory in particular. While individuals with FTD outperformed those with AD on tests of memory, those with AD outperformed those with FTD on tasks of executive functioning, mirroring prior results. Overall, no differences were found between the 3 groups in confrontation naming, recognition memory or basic attention. However, Pachana (2006) suggests that further examination of scores across cognitive domains, in addition to interpretation of individual neuropsychological scores, may be useful in future research.
Rakovsky (2002), using multivariate analysis of variance, also revealed that patients with FTD performed significantly worse than patients with AD on tests of executive function (such as letter and category fluency) but significantly better than those with AD on tests of memory (such as block design and the clock drawing test). Overall, the FTD patients were more impaired on tasks that are sensitive to frontal lobe dysfunction (eg. word generation, verbal fluency), and less impaired on tests sensitive to dysfunction in the medial-temporal and parietal cortices (eg. memory and visuo-spatial abilities), more commonly associated with AD. Overall this study reflects upon previous findings and further demonstrates similarities and differences across these two forms of dementia in relation to age related controls.

In summary, prior research acknowledges the cognitive decline associated with FTD, its differences from other forms of dementia such as AD and also its similarities. Research done by Braaten (2006), Pachana (1996) and Rakovsky (2002) clearly demonstrate that while less prevalent, FTD is in fact comparable to other forms of dementia in terms of psychological impairment and symptom severity. What these studies fail to do is further assess and acknowledge each form of dementia differentially and furthermore, relate those results to real life activities and performance. The current study aims to expand upon these findings by focusing on FTD specifically, by assessing how symptom patterns and performance are presented in this particular population, and furthermore, by then assessing the relationship between neuropsychological dysfunction and individual abilities to perform daily life skills.
Activities of Daily Living (ADLs) in FTD

ADLs: What are They/Why are They Important?

A major area of study at this time is how the symptoms and cognitive deficits associated with dementia affect an individual’s ability to function in everyday life (activities of daily living). As recognized by the U.S. Department of Health and Human Services (HHS, 1990), ADLs are understood to include actions such as eating, bathing, dressing and toileting. Other, more complex forms of ADLs include communication, being aware of time and space, orientation and daily tasks that rely on memory, such as shopping. As emphasized by the HHS (1990), ADL’s can significantly predict various actions and outcomes in a person’s life.

The inability to perform ADLs can predict admission to a nursing home or other non-independent living arrangements and may affect both hospital and physician services. With a significant prevalence in demented populations, ADLs are now considered as a standard measure of care and are tied into patient evaluations and long term care (Wiener et. al, 1990). Additionally, individual abilities regarding ADLs now predict insurance coverage and help to shape future policies (Wiener et. al, 1990). Overall, these findings highlight the importance of accurately assessing and understanding ADL performance, particularly in elderly and demented populations, who according to the HHS, are most susceptible to ADL disturbances.
Measuring ADLs

The way in which ADLs are measured varies from one study to the next. However, there are essentially two methods for assessing ADLs: questionnaires completed by patients or informants (typically caregivers) or direct observation measures.

In terms of informant-rated measures, although there is some consensus across surveys as to which activities should be included, there is a great deal of variation in the way surveys ask about ADL functioning (HHS, 1990). Since informant- or self-report instruments do not incorporate strict definitions for the activities being assessed, or for the possible response categories, estimates may vary simply because respondents interpret the questions differently. Furthermore, there may be a tendency for patients to underestimate their disability when filling out a questionnaire or, for a caregiver’s level of burden to impact their perception of ability/disability in the patient they are completing the ADL rating about.

In contrast, observation-based measures occur in real time and differ in that they require the patient to act out several ADL tasks directly in front of an unbiased examiner. Rather than relying on the memory or prior observations of a caretaker, direct observation tests can create real life experiences in a setting where performance can be more objectively assessed.

ADLs in FTD

One study by Bouwens (2009) used the Blessed Dementia scale to demonstrate how ADLs have a direct relation with cognitive decline in dementia patients. Within this study
specifically, where N=615, significant correlations were found between cognitive decline and impairment in ADL’s across various types of dementia. The strongest correlations were found for those with Frontotemporal Dementia (-0.80) and dementia of the Alzheimer’s type (-0.60) with moderate associations for Vascular Dementia, (-0.50), and Parkinson’s disease (-0.050).

Results of Bouwens (2009) suggest that different types of dementia are characterized by a specific pattern of cognitive functioning which then reflect varying abilities in daily life functioning. While this study provided a generalized relationship between FTD and ADL performance, one weakness in this study was the use of the Blessed Dementia Scale (BDS), which is an informant-based measure. While the BDS assesses specific forms of functioning (eg. ability to perform household tasks, manage money etc.) it requires the informant to use their own interpretation and memory for assessment, which as previously mentioned, leaves room for error. A specific study by Cole (1990) found the inter-rater reliability on the BDS to be particularly low, indicating that when used by multiple raters, they were unlikely to give similar scores for the same individual.

Further research by Kipps et al. (2009) assessed social and emotional processing of individuals with Alzheimer’s disease and FTD, in relation to ADL performance. By use of the Disability Assessment for Dementia of Activities in Daily living (DAD-ADL) and the Cambridge Behavioral Inventory (CBI), both caregiver-based assessments, these researchers found that the FTD group had significantly greater difficulty in performing ADLs as measured by the DAD-ADL, and had higher scores on the CBI compared to the AD group (indicating greater dysfunction). Though no relationship was found between
social and emotional processing and ADLs, there was a significant difference in ADL performance between groups. Furthermore, ADL ratings were significantly correlated with caregiver apathy, suggesting that ratings were subject to personal bias, again creating a potential confound (Kipps et. al, 2009).

Mioshi et. al (2007) interviewed fifty nine individuals with FTD and AD, along with their caregivers, in order to assess the relationship between cognitive abilities and activities of daily living. Similar to Kipps et. al. (2009) caregivers were interviewed with the DAD-ADL, where patients were also rated on the Clinical Dementia Rating Scale (CDR) and the Addenbrooke's Cognitive Examination-Revised scale (ACE-R) for neuropsychological assessment. In comparison to those with AD, results demonstrated that those with Picks Disease (or the behavioral variant of FTD) performed the worst on the DAD-ADL, whereas those with Progressive Non-Fluent Aphasia (PNFA) or semantic dementia (the language variant of FTD), were least impaired, with AD ranging in the middle.

In contrast to prior findings, ADL performance did not directly correlate with cognitive decline. Results from Mioshi et. al. (2007) found that groups who demonstrated the best ADL performance (PNFA/Semantic Dementia) showed lower cognitive functioning and vice versa. As to be expected, scores on the DAD-ADL did not correlate with those from the ACE-R, again contradicting previous results. Further analyses did demonstrate a unique pattern of deficits for each group, supporting prior studies, but again, were limited by their use of the DAD-ADL, subject to informant bias and flaw.
Lastly, results by Razani et. al (2007), using an observation-based ADL task, do support previous findings in that cognitive functioning was significantly correlated with ADL performance in patients with dementia (which included patients with FTD). Further results demonstrated that this relationship was strongest for tests of executive functioning, including verbal fluency, a complex test of cognitive flexibility and reasoning (recognized by Rakovsky (2002) as areas of weakness for those with FTD). Overall, findings from this study suggest that there is a direct relationship between cognitive dysfunction and ADL performance in those with FTD, but fails to detail this relationship specifically.

Results from Razani et. al (2007) support and furthermore, improve upon other studies by using the Direct Assessment of Functional Status (DAFS), an observation based measure of ADL performance. Unlike the BDS or the DAD-ADL, research by Zanetti et. al, (1997) assessed the validity of the DAFS, and found it to correlate significantly with a brief cognitive measure, with a nonverbal measure of intelligence and a clinical dementia rating in 93 patient with AD. The authors concluded that this is a valid measure that not only informs about levels of disability, but also the state of dementia illness. Based on these findings, it is this measure of ADL performance that will be used in the present study.

In sum, previous research by Mioshi et al. (2007), Kipps et. al. (2009), and Bowens (2009) demonstrates a clear association between cognitive deficits and a decline in ADL, but failed to use an accurate measure of ADL performance. Results from Razani et. al (2007) identified similar patterns and made use of a more objective ADL measure, but
did not recognize details specific to FTD. While these studies did well to demonstrate a
general relationship between cognitive decline and ADL function, results varied across
symptom patterns, dementia subtypes and ADLs measured. Each study identified specific
variables, but failed to elicit an overall picture relevant to FTD specifically; it is with this
in mind that the current study is being done.

Specific Purpose/ Aims of the Current Study

The current study aims to improve upon prior research by utilizing an observation-based
ADL task to specifically assess patterns of cognitive and ADL performance in patients
with FTD. In analyzing patterns of performance on these tasks, the current study hopes to
better depict how the cognitive decline associated with FTD manifests in daily life
functioning.

Aim #1: To better characterize patterns of deficits in specific
neuropsychological/cognitive and ADL domains in patients with FTD, where the
independent variable is the actual test score and the dependent variable is the pattern in
relation to normal controls (either higher, lower or no difference) on the two separate
types of test (ADL and cognitive).

Hypothesis for Aim #1: Based on previous literature, we hypothesize that the FTD group
will show specific patterns of performance on cognitive domains. Specifically, they will
have greater deficits in the domains of executive functioning and memory.
Aim #2: To assess specific patterns of performance on ADL subscales/skills in patients with FTD.

Hypothesis for Aim #2: We hypothesize that while FTD patients will perform poorly on all aspects of daily functioning, performance will be worse on activities that rely heavily on executive and memory skills.
Method

Participants

Approximately 70 individuals were used from a larger database. The data was collected over an 11 year period as part of a larger, longitudinal study, designed to assess cognitive functioning, activities of daily living, and caregiver burden in patients with various forms of dementia. For this study, 13 patients with Frontotemporal Dementia and 57 age-matched controls (56-69 years) were used. FTD patients were recruited from the Harbor-UCLA Medical Center and the UCLA Alzheimer’s Disease Center (ADC). The diagnosis of FTD was predetermined by behavioral neurologists and neuropsychologists at the UCLA ADC and/or the Geriatric Neurobehavior Clinic at the West Los Angeles Veteran’s Administration Medical Center.

Both sites used the Neary et al. criteria (Neary et al., 1998) in making their diagnosis of FTD. The Neary criterion, established in 1998 incorporates three clinical syndromes used to identify FTD. This includes early decline in social and personal conduct, emotional blunting and loss of insight. Another core feature included impairment in regulation of personal conduct among supportive diagnostic criteria: behavior dysfunction, speech and language impairment and physical signs. Other measures including neuroimaging were also utilized in identify FTD. Lastly, FTD is primarily known and identified based on its insidious onset and gradual progression.

For the purpose of this study, normal, healthy individuals were either caregivers of patients or healthy controls recruited from the greater Los Angeles community, by various means of advertisements. In order to determine these individuals as appropriately
healthy/normal, a thorough medical history was gathered to screen out those with major medical conditions known to affect cognitive ability. As such, those with a history of head injuries, major affective or psychotic disorders, seizures or other neurological disorders and substance abuse were excluded from the study.

Demographically, there were a total of 13 individuals in the FTD group, with 9 males and 4 females. Within this group, 3 had attended at least 12 years of school, with 3 more attending at least 13, 1 at 14 years of education, 1 at 18 and 1 at 22 years.

There were 8 FTD participants who identified themselves as Caucasian Non-Hispanic, 1 identified as Hispanic, 1, African American and 1 who identified as being of Asian descent. Only 10 of 13 reported a primary language, where 9 noted English to be their primary language and one reported being a native non-English speaker. Only 3 of the participants reported prior medical conditions, where 4 reported none and 6 did not reply. In addition to FTD, 5 identified the presence of other neurologic problems, 4 had a family history of dementia and 4 also reported prior head injuries.

Within the FTD group, 3 had received prior psychiatric treatment, 2 reported prior substance abuse and 5 were currently on medication.

There were a total of 57 individuals in the NC group with 11 males and 46 females. Of the participants in this group, 10 had attended at least 12 years of school, with 9 more attending at least 13, 11 at 14 years of education, 1 at 15, 13 at 16 years and 9 at 17 or more years.
There were 44 who identified themselves as Caucasian Non-Hispanic, 7 who identified as Hispanic, 3 as African American, 1 who identified as being of Asian descent and 2 who reported under the “other” category. Of the total participants in this group, 50 used English as their primary language and 7 reported being a native non-English speaker.

Thirty-three of the participants reported prior medical conditions, where 20 reported none and 4 did not reply. In addition to FTD, only 1 identified the presence of other neurologic problems, 13 had a family history of dementia and 6 also reported prior head injuries.

Within the NC group, 11 had received prior psychiatric treatment, 2 reported prior substance abuse and 46 were currently on medication.

Both groups completed a neuropsychological test battery which took approximately 2 - 2 ½ hours to complete, and a 30 minute direct observation task to measure ADL performance. Overall, testing was divided into two or more sessions depending on the availability, time and needs of the participants. At the end of each testing interval (both baseline and follow up sessions), patients in both groups were paid for their participation.

**Measures Administered**

**Activities of Daily Living**

**Direct Assessment of Functional Status (DAFS)**

The DAFS is a standardized, observation-based test which assesses the ability to perform skills of daily living (Loewenstein et al., 1989). Intended for older individuals, it has
proven useful in indicating the level of functional capabilities for those with Alzheimer's disease, dementia and schizophrenia (Johnson et. al, 2004).

The DAFS is composed of seven ADL ability areas, where specific tasks are administered in order to assess these (7) core functional domains. Such tasks are simulated to represent real life situations, thereby aiding in the validity of this measure (Johnson et al. 2004). The first area measured is orientation, assessing the ability to tell time using a clock and further demonstrates orientation to person, place and date. These aspects are measured through two subscales, Orientation to Time and Orientation to Date. The second domain, communication skills, measures a patient’s communication skills, their ability to use a telephone and their ability to prepare and mail a letter (subscales Using a Phone and Letter Writing). The third component, transportation skills, assesses the ability to identify road signs and knowing driving rules (Subscales Identifying Traffic Signs and Driving Rules). The fourth area, financial skills, assess the ability to identify currency, the ability to count currency, the ability to write a check and the ability to balance a checkbook (subscales Identifying Currency, Counting Currency, Writing a Check and Balancing a Checkbook).

The fifth area, shopping skills assesses the ability to follow a grocery list directly as well as indirectly, via free recall of items needed/listed. This subscale also measures the ability to “shop” by recognizing items and the ability to make correct change. These abilities are represented in the subscales labeled Shopping Free Recall, Shopping Recognition, Shopping with a List and Making Correct Change. The sixth and seventh subscales include grooming and eating. Grooming abilities demonstrate the ability to perform
certain grooming skills that are considered to be basic, including the ability to comb hair and the ability to use a toothbrush. The eating subscale measures the ability to perform eating tasks, such as using utensils correctly (Williams-Gary, 2013).

Overall, the points received in each domain are tallied to obtain a total DAFS score. For the purposes of this study, all domain scores were also analyzed individually, with the exception of Eating and Grooming subscales, since most patients will obtain a perfect score on these measures.

Neuropsychological Tests
The following neuropsychological tests were selected to be sure that major cognitive domains were assessed:

California Verbal Learning Test- II Short-Form (CVLT-II SF)
The California Verbal Learning Test- II Short-Form measures verbal learning and memory (Delis et. al, 1999). More specifically, the CVLT-II SF measures various encoding strategies, learning rates, error types and other forms of processing. These include general and working memory retrieval, retroactive inference and chunking (Delis et. al, 1999). Furthermore, the CVLT-II also includes forced-choice items useful for detecting malingering and has been found to be a strong predictor of pre-clinical Alzheimer’s disease (Delis et. al, 1999).

Similar to the full version, this form requires participants to learn and remember a single list of words, in this case a list of 9 words, across four different learning trials. The words each belong to one of three categories. The initial trial or learning trial occurs when the examiner reads the list of 9 words to the participants and asks them to
immediately recall as many as they can. After a 10 minute (long-delay) break, in which the participants complete other nonverbal tests, they are asked to freely recall the 9 items again (free recall condition), then are given cues to recall the list (cued recall condition) and finally asked to recognize the list items which are embedded among foils (discrimination condition) (Delis et. al, 1999).

Scoring of the CVLT-II was done by using scoring software which allows for accurate data entry and score reporting, along with in-depth information and more detailed analyses if needed (Delis, Kramer, Kaplan & Ober, 1999). For the purpose of this study, we only examined a % savings score, which was calculated in the following way: number of items recalled during the 4th learning trial divided by number of items recalled during the free recall condition, then multiplied by 100.

**Rey-Osterrieth Complex Figure (ROCF)**

The Rey Osterrieth Complex Figure or ROCF is a two dimensional test of non-verbal memory, with specific measures in visual-spatial/constructional memory and visual memory. It consists of a singular drawing with 18 details, among which, are a variety of shapes including squares and triangles, to circles and crosses, arranged in a specific order around a central rectangle (Mitrushina et al, 2005).

In order to complete this task, the participant is instructed to carefully copy the given design with pencil onto a separate piece of paper, placed to the side of the drawing. For the memory component, 30 minutes later, they are asked to draw the design again from memory.
For the purpose of the current study the (standard) Meyer & Meyer (1992) scoring criteria was used. Overall, the primary scoring criteria is based on an assessment of 18 particular characteristics of the figure. For each of the 18 areas, a possible score of 0 to 2 points is possible. Points are based on the correct placement and construction of each item and are rewarded based on the given criteria, for a total possible score of 0-36. Two points are given when an item is placed and reproduced correctly; 1 point is given for correct reproduction, but wrong placement, 1 point is given for correct placement but distorted reproduction, and 0.5 point is given if an item is incomplete but recognizable. The criteria for which a zero score is received requires that the item is absent or not recognizable.

For the purpose of this study a % savings score was used where \( \% \text{ Sav} = (\text{30 minute delay/ initial copy score}) \times 100 \).

**The Wisconsin Card Sorting Test (WCST)**

The WCST is a test of executive functioning that assesses cognitive flexibility, and abstract thinking (Mitrushina et. al, 2005). For this study, the 64 card version was used. For the WCST individuals are presented with a deck of 64 stimulus cards, which they must match to four designated response or reference cards placed in front of them. They are given very little instructions and the object of the task is for participants to sort the cards based on three different principles: color, shape/form and number (Mitrushina et. al, 2005). They are given minimal feedback of “correct” or “incorrect from the test administrator. In order to obtain one categorical sort, the participant needs to obtain 10 consecutive correct sorts. Once they have obtained the category, the principal to which
they are to sort switches. This task is continued until the participant has sorted all 64 cards. A number of different outcome measures are obtained. Table 1 presents the outcome scores that were used for this study.

Table 1

*Primary Outcome Measures for the Wisconsin Card Sorting Test*

1. **Categories Completed**: refers to the total number of correct principles sorted, where credit is earned for the completion of 10 consecutive correct matches.

2. **Percent Conceptual Level Responses**: measures the participant’s insight into the correct sorting principle and based on consecutively correct responses in the first run of three or more.

3. **Total Errors**: measures the number of sorting errors made throughout the task

*Wechsler Adult Intelligence Scale (WAIS-IV) – Digit Span*

The WAIS-III Digit Span test assesses short-term auditory memory and attention. This test can be broken down into two basic components, the digit span forward and backward. The “forward” test is composed of random number sequences which are read off by the test administrator and then repeated back by the examinee in the exact same order. For each number sequence repeated correctly a point is given towards the total, and the amount of numbers in the sequence increase, becoming longer and more challenging. In the “backward” version, the task is the same with the exception that the examinee must repeat the random number sequence in reverse, starting with the last number they heard and working backward. Again, with each correct response a point is given and the
sequence becomes gradually more complex until three scores of zero are consecutively reached; at this time the test is concluded (Wechsler, 2008).

**FAS**

The FAS is a test of language proficiency where participants are asked to freely recall responses based on instructed categories. Participants go through four stimulus phases. The first is letter F, the second A, the third S and the fourth Animals. In the first phase, participants are asked to recall as many words as possible beginning with the letter “F” until 60 seconds has passed. The same occurs in the second phase, except participants are instructed to recall words beginning with the letter “A.” The same is true of the third phase, where participants recall word beginning with the letter “S.” In the fourth and final phase participants are asked to think of as many animals as they can, again, within a 60 second time frame. Each phase is done consecutively, without significant or intended delay between each phase.

Participants scores are based on the number of words stated in each phase correctly ie. those that follow from the instructions and correlate correctly to the given stimulus (F,A,S or Animals). Each phase is broken down into 15 second quadrants under which words are scaled for each section (four in total) per phase.

**Data Analysis**

Z-scores were created for the neuropsychological variables in order to put all measures on a comparable scale. Mean scores for each neuropsychological variable for the FTD
patients were converted into z-scores by subtracting the score of each patient from the mean of the normal control scores and dividing the difference by the standard deviation of the normal controls. Z-scores were then used as dependent variables for comparison of the performance of the FTD patients across the various neuropsychological outcome measures. It should be noted that z-scores were adapted so that the greater the value the better the performance.

To further assess specific abilities within each group, the percentage of correct responses was calculated for each of the 14 DAFS subscales for FTD patients and normal controls. The calculations were conducted by taking individual scores on a given subscale and dividing it by the total possible score for that subscale, and then multiplying that by 100. Percentage scores from the FTD group were compared to those of the normal control patients to better assess similarities and differences in their performance patterns.

Due to the multiple comparisons, the p-value required for statistical significance was lowered to .01 rather than the standard .05 value. Significance values for the follow-up analyses were set at the standard .05 level. While we recognize that this may not entirely protect against Type I error, a more stringent criteria would increase Type II error due to the small sample size.
Results

A repeated measures ANOVA assessing the neuropsychological $z$-scores of the FTD group revealed no significant differences, $F(1, 11) = 6.031, p > .05$. A repeated measures ANOVA assessing the DAFS $z$-scores of the FTD group revealed significant differences, $F(1, 11) = 11.533, p < .05$.

To assess percentage scores, a mixed design ANOVA, using groups as the IV, and the 14 DAFS subscales as the DVs, showed a significant interaction between groups and DAFS subscales $F(13, 832) = 47.56, p < .001$. Follow-up one-way ANOVAs and post hoc comparisons were conducted. Table 2 presents the results of the one-way ANOVA comparing the FTD and normal control groups on each DAFS subscales individually. It appears that the normal controls outperformed the FTD group on all DAFS subscales with the exception of the Check Writing/Writing a Check subscale.

To examine further differences within the FTD group, a one-way repeated measures ANOVA was used to analyze the patterns of percentage scores on the DAFS subscales. Results revealed significant differences within the FTD group, $F(13, 143) = 10.81, p < .001$. Post Hoc analyses revealed that the FTD group performed the same on DAFS subscales of Balancing a Checkbook, Shopping Free Recall, Shopping Recognition, and the Shopping subscale, making correct change, but that performance was worse on these subscales relative to all of the others ($p$ values < .05; see Figure 1).

To account for DAFS subscale differences within the NC group, a second repeated measures ANOVA was performed. Results revealed a significant difference between DAFS subscales within the normal controls group, $F(13, 689) = 38.85, p < .0001$ (see
Figure 2). The normal controls performed similarly on the DAFS subscales of balancing a checkbook and shopping recognition in relation to all other DAFS subscales. Additionally, the scores showed that the shopping free recall scores were significantly lower than all other subscales as well. Lastly, results showed that scores for shopping with a list, orientation to date and identifying currency, were significantly greater than those for writing a check, using a phone and writing a letter. The ability to read traffic signs, comprehend driving rules and count currency were also greater than the ability to use a phone or write a letter, but not different than the ability to write a check.

It should be noted that while the pattern of differences in DAFS subscale performance is similar in both groups, those with FTD displayed far more impaired performance as compared to the normal controls, and the magnitude of the differences in subscale performance was greater in the FTD group.
Table 2

*One-Way ANOVAs comparing DAFS performance of FTD and controls*

<table>
<thead>
<tr>
<th>DAFS Subscale</th>
<th>df</th>
<th>F</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>% Orientation to Time</td>
<td>1, 64</td>
<td>6.974</td>
<td>.010</td>
</tr>
<tr>
<td>% Orientation to Date</td>
<td>1, 64</td>
<td>17.295</td>
<td>.000</td>
</tr>
<tr>
<td>% Telephone Skills</td>
<td>1, 64</td>
<td>7.984</td>
<td>.006</td>
</tr>
<tr>
<td>% Letter Writing</td>
<td>1, 64</td>
<td>12.030</td>
<td>.001</td>
</tr>
<tr>
<td>% Identifying Traffic Signs</td>
<td>1, 64</td>
<td>30.791</td>
<td>.000</td>
</tr>
<tr>
<td>% Driving Rules</td>
<td>1, 64</td>
<td>17.071</td>
<td>.000</td>
</tr>
<tr>
<td>% Counting Currency</td>
<td>1, 64</td>
<td>14.958</td>
<td>.000</td>
</tr>
<tr>
<td>% Writing a Check</td>
<td>1, 64</td>
<td>2.637</td>
<td>.109</td>
</tr>
<tr>
<td>% Balancing A Checkbook</td>
<td>1, 64</td>
<td>13.711</td>
<td>.000</td>
</tr>
<tr>
<td>% Identifying Currency</td>
<td>1, 64</td>
<td>4.760</td>
<td>.033</td>
</tr>
<tr>
<td>% Shopping Free Recall</td>
<td>1, 64</td>
<td>50.505</td>
<td>.000</td>
</tr>
<tr>
<td>% Shopping Recognition</td>
<td>1, 64</td>
<td>66.399</td>
<td>.000</td>
</tr>
<tr>
<td>% Shopping with a List</td>
<td>1, 64</td>
<td>9.241</td>
<td>.003</td>
</tr>
<tr>
<td>% Making Correct Change</td>
<td>1, 64</td>
<td>51.424</td>
<td>.000</td>
</tr>
</tbody>
</table>
Figure 1. FTD Percentage Scores Across DAFS Subscales

Figure 2. Normal Control Percentage Scores Across DAFS subscales
Discussion

Previous research has demonstrated that those with FTD have deficits in performing neuropsychological tests when compared to normal controls. However, only a few studies have been conducted with FTD, examining daily functional abilities. Furthermore, studies examining ADLs in FTD have not evaluated specific patterns of deficits in the daily tasks they are likely display. The current study aimed to assess the specific types and severity of deficits FTD patients are likely to show on a performance-based ADL test. The current study intended to evaluate these patterns of daily dysfunction using a performance-based task in order to further understand the effects of this disorder on the ability of patients to function independently.

Neuropsychological Performance

Initial analyses confirmed the findings of Braaten (2006), Pachana (2006) and Rakovsky (2002), in that those with FTD performed significantly worse on all measures of neuropsychological ability in comparison to normal controls (as demonstrated in their z-score averages). However, there was no specific pattern of neuropsychological test performance for FTD patients (assessed via Z scores). That is, the FTD group comparisons of the neuropsychological z-scores revealed that there were no significant differences in performance between the various neuropsychological measures. Our current results in fact, are inconsistent with some previous findings. Braaten (2006) and Rakovsky (2002) found that individuals with FTD performed poorly on task of executive functioning and verbal and non-verbal memory as also mentioned by Pachana (2006). In our study we found performance on tasks such as visual/auditory memory, recognition
memory, language proficiency, spatial/constructional memory, recall and basic attention to be similarly impaired. It is unclear why our study did not find distinct patterns of deficits on neuropsychological tests for FTD patients. It is possible that the use of z-scores for standardizing scores or the small sample size did not allow for detection of subtle differences in performance across the measures.

**ADL Performance**

In assessing ADL performance, impairment was significantly greater for those with FTD in relation to normal controls. This supports the findings of Kipps et. al. (2009), who also found that those with FTD were outperformed by age matched controls, in addition to those with Alzheimer’s disease (AD), suggesting that these individuals are most subject to deficits in ADL performance within these groups. Results from the present study help to verify the deficits specific to FTD and thereby expand upon prior findings by further detailing what areas are even more specific to this population. Not only was it confirmed that normal controls outperformed those with FTD, but results make it evident that there are deficits within the FTD population which further differentiate this population from normal controls as well as those with Alzheimer’s Disease.

The percentage score analyses clearly demonstrated that there are in fact areas of daily functioning in which those with FTD perform better or worse on, in relation to their overall ADL abilities. The primary deficits were found in tasks related to shopping and financial skills, with specific deficits in shopping recognition and free recall, writing a check and calculating correct change. Comparisons of the percentage scores suggests that these abilities are particularly impaired in those with FTD, compared to other ADL tasks,
such as those related to transportation and orientation skills. As reported by Razani et. al. (2007) and Razani et al. (2010), these tasks appear to be directly related to the cognitive domains of executive functioning and memory. The primary damage in FTD is in the areas of frontal lobe and temporal lobes, known to govern executive functions and memory, thus it is not surprising to find that performance on these tasks are most affected.

Results suggest that those with FTD may neglect daily responsibilities related to performing complex tasks, particularly those requiring memory, planning, and organization. Overall, these findings indicate that there may be a need for caregiver assistance in performing tasks such as shopping and financial management. Specific to the DAFS subscales listed, task regarding item recognition/identification in general may be difficult for these individuals, they may have trouble recognizing what may have once been familiar items and may have trouble recalling previously learned information. In the context of this study, these skills directly affected the ability to shop and work with money, though future studies may look at these abilities across other contexts in which they may be used. The inability to write a check and calculate correct change again, supports the inability to perform executive functions, mental organization and processing. Overall these patterns are directly reflective of the skills assessed, support the findings of prior research, and also expand upon them by detailing specific areas of impairment for those with FTD.

Interestingly, the normal control group displayed a similar pattern of performance, in which they did worse on the shopping recognition, shopping free recall and task of
writing a check, in relation to all other abilities. However, their deficits on these tasks were not nearly as great as those with FTD and differences between these subscales and the others were not nearly as large as those found in the FTD group. This suggests that 1) the normal control group also has deficits in daily tasks that require use of executive and memory functioning (perhaps due to deterioration of brain regions as a function of natural aging), but that 2) their deficits are not “pathological” or disease related as they are in the FTD group. Nonetheless, deficits in everyday functioning, no matter how mild in healthy controls can be important to document (Cahn-Weiner, Boyle, & Malloy, 2002). As previously mentioned, the presence of ADL impairment significantly affects the ability to be admitted to nursing home or receive other healthcare benefits. For example, Jackson and Burwell (1989) found that the level of ADL disability in patients with dementia (as well as their caregivers) may be helpful in determining long term care needed for the patient.
Summary

Overall, results from this study support previous research and also expanded upon it by utilizing multiple comparisons and analyses. It is clear that those with FTD perform worse on both cognitive and ADL tasks relative to their normal controls. What we now know, however, is that in terms of ADL performance, there are in fact tasks pose more difficulty for FTD patients.

Limitations

Overall this study proved to hold significant findings but was not without flaws. One of the major areas future studies may improve upon is the number of participants in the experimental group. While 13 is considered a large sample by standard for FTD, future studies may improve upon this research by accommodating a larger sample size where possible.

Furthermore, the difference in sample size may have had an effect on analyses (57 for NC and 13 for FTD) as could have the gender ratio (Female>Male) which was not accounted for in this study. Other demographic variables such as race, ethnicity, education and other factors previously mentioned could be controlled for or analyzed as a covariate to account for effects of these variables as well. Overall, assessing cultural or gender differences may prove relevant for future research.

Another factor this study did not control for was the different forms of FTD, the behavioral variant vs. the language variant, which manifest differently and as such may affect the ability to perform ADL tasks separately.
**Future Research**

Future studies would do well to further compare the relationships of neuropsychological abilities and ADL performance as done by Bowens et. al (2009), but accommodate and account for the relational patterns of performance, as opposed to just documenting areas of abnormality.

Future studies may also expand upon the current findings by comparing patterns of ADL performance across other forms of dementia, or as mentioned, within the separate subtypes of FTD. For an even more detailed analysis, performance patterns could be assessed in comparison to the three areas of deficits identified by the National Institute of Aging (NIA, 2013), including progressive behavior/personality decline, progressive language decline and progressive motor decline.

To better assess the utility of the DAFS vs. other mentioned ADL measures (Blessed Dementia Scale (BDS) etc.), future researchers would do well to compare patterns of performance within a single group across different ADL tests. This would prove useful in further assessing patterns of performance more accurately and also allow for comparisons between what is observed and/or revealed by caregivers and what is objectively identified. This could then be related to caregiver biases and expectations as well as caregiver burden.
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