

A thesis submitted to the University of Birmingham for the degree of
Doctor of Clinical Psychology (Clin.Psy.D.)

VOLUME I

RESEARCH COMPONENT

Literature Review

Frontal-variant Frontotemporal Dementia and ‘Theory
of Mind’ Tasks:
A Systematic Review of the Research

Research Paper

The Family Experience of Frontal-variant
Frontotemporal Dementia:
A Qualitative Study

by

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Overview

This thesis is submitted in partial fulfilment for the requirements of the degree of Doctor of Clinical Psychology at the School of Psychology, University of Birmingham. It comprises of a research and a clinical volume.

Volume I

Volume I is the research component of the thesis. It consists of two papers, the first of which is a review of the literature that uses ‘Theory of Mind’ (ToM) tasks with people with frontal-variant frontotemporal dementia (fvFTD). All the research identified is systematically appraised in terms of the methodology and the quality of the published reports. The evidence indicates that ToM is impaired in people with fvFTD; however more robust findings were evident from research which comprehensively measured neuropsychological functioning and used established and well known ToM tests. More recent research has diversified into exploring other aspects of social cognition, such as emotional processing and empathy, and their relationships with ToM. The nominated journal for this review paper is ‘Neuropsychologia’.

The second paper is a qualitative research project that explores the experiences of family members of people living with fvFTD. The research questions were: How does the development of fvFTD in a working age person affect the family experience of living with that person, and how might mental health services respond to the needs of those family members? Individual in-depth interviews were carried out with six relatives (including partners, a sibling, and an adult child), and Interpretative Phenomenological Analysis (IPA) was used to analyse the data resulting in the emergence of four main themes. The findings demonstrate how family caregivers of people with fvFTD have to contend with specific behavioural challenges and personality changes associated with the condition. The study also indicates that knowledge about fvFTD is lacking in both carers and professionals alike, causing uncertainty and long periods waiting for a diagnosis, which adds to the burden of care for these people. Services need to be developed to cater for specific individual needs and awareness needs to be raised in all health care services. The nominated journal for this research paper is ‘Dementia: The International Journal of Social Research and Practice’.

Volume II

Volume II is the clinical component of the thesis, which consists of five clinical practice reports (CPRs) that describe and evaluate clinical work carried out during clinical placements throughout the training course. The first CPR ‘Psychological Models’ formulates the case of an 18-year-old woman with anxiety symptoms from a systemic and a cognitive perspective. The second CPR ‘Small Scale Service-Related Research Project’ is a qualitative evaluation of a drop-in service for young people leaving care. The third CPR ‘Single Case Experimental Design’ evaluates the intervention designed to support a woman with a moderate learning disability and behaviour that challenged services. The fourth CPR ‘Case Study’ details the neuropsychological assessment of an 81-year-old man with memory problems. The fifth CPR was presented orally and it describes the use of Cognitive Analytic Therapy with a woman presenting with anxiety following treatment for breast cancer. The abstract is included here only.

NB: Throughout both Volumes I & II all identifying information relating to clinical clients and research participants has been changed in the interests of their anonymity and confidentiality.

Dedication

Firstly, I would like to dedicate this thesis to my parents and family, to my friends and fellow trainees, who have all been there for me in their own individual ways, helping me through the journey of clinical training; my heartfelt thanks to you all.

I would also like to dedicate this work to my mentor and leader in faith, Daisaku Ikeda, who is president of the Soka Gakkai International (SGI), a Buddhist organisation dedicated to the promotion of peace through the Buddhist values of courage, wisdom, and compassion.

By extension I would therefore also include all SGI-UK members, and particularly my fellow practitioners in the local area: the 'Heart of England' headquarters.

NMRK

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Birmingham, May 2009

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Literature Review

Frontal-variant Frontotemporal Dementia and ‘Theory of Mind’ Tasks: A Systematic Review of the Research

ABSTRACT

The purpose of this review was to systematically evaluate the research evidence indicating that 'Theory of Mind' (ToM) is defective in people with frontal-variant frontotemporal dementia (fvFTD). A literature search was carried out on the Psychinfo, Medline, and Embase online databases to identify all the literature in the past ten years using ToM tasks with people with fvFTD. Nine relevant papers were identified since the year 2001 and included in this review. The search also uncovered a paper by Kipps and Hodges (2006) which was a conceptual analysis of the relationship between fvFTD and ToM. All the studies mentioned in that work are reviewed here. This paper is a more systematic appraisal of the methods used and the quality of the published reports. More robust findings were evident from research which comprehensively measured neuropsychological functioning and used established and well known ToM tests. More recent research has diversified into exploring other aspects of social cognition, such as emotional processing and empathy, and their relationships with ToM. The evidence indicates that ToM is impaired in people with fvFTD.

INTRODUCTION

Frontal-Variant Frontotemporal Dementia

In frontotemporal dementia (FTD), the frontal and temporal areas of the brain are subject to degeneration, and this gives rise to characteristic patterns of symptoms. Degeneration predominantly in the temporal lobes is called temporal-variant FTD (tvFTD) and primarily results in language difficulties. In contrast people with frontal-variant FTD (fvFTD), where degeneration is located primarily in the frontal lobes, present with: insidious changes in personality and behavioural disturbances; interpersonal difficulties characterised by a lack of empathy or concern for others; disinhibition or other socially inappropriate behaviours; and a general lack of insight and apathy (The Lund and Manchester Groups, 1994; Appendix 2). People with fvFTD generally present in their 50s and 60s with these symptoms. They are not generally amnesic, as would be common in other early onset dementias (e.g. Alzheimer's disease), and in fact they often perform within the normal range on memory tests in the early stages (Cycyk & Wright, 2007). This, alongside their unusual behaviours, can cause difficulties with diagnosis and often lead to erroneous psychiatric diagnoses (e.g. depression, schizophrenia).

Social Functioning, Empathy & FvFTD

People with fvFTD typically lack insight into the changes in their own behaviour and appear oblivious to the effects that these have on others. These early changes in social behaviour may reflect a difficulty in the perception of emotions and emotional cues (Kosmidis, Aretouli, Bozikas, Giannakou, & Ioannidis, 2008), including emotional states inferred from facial and/or vocal expression; and responses to body gestures (or paralinguistic cues; Lough *et al.*, 2006). Decety and Jackson (2004) proposed a framework for understanding empathy, which is relevant to the deficits seen in fvFTD. In their proposal they suggest that empathy has three broad components: an affective response to another person, usually sharing their emotional state; a cognitive capacity, to take another person's perspective; and a regulatory or inhibitory aspect, in order to take account of the self and others' feelings (Kipps & Hodges, 2006).

The Functional Anatomy of FvFTD

Neuro-imaging and pathological studies of people with fvFTD have indicated severe atrophy of the frontal and anterior temporal neo-cortex, which may be mostly centred in the orbital regions or, with the later progression of the disease, atrophy may extend further into the anterior cingulate and dorsolateral frontal cortex (Diehl *et al.*, 2004). Furthermore, imaging studies have implicated the ventromedial prefrontal cortex as a critical region affected in these conditions (Williams, Nestor, & Hodges, 2005). Notably, this is an area also thought to be involved in social functioning and particularly to be the neural basis for the representation of mental states (Frith & Frith, 2003). So in summary, the networks of structures susceptible to damage in fvFTD are the frontal lobes, the orbital, medial and dorsolateral prefrontal cortex, and the temporal poles.

‘Theory of Mind’

In order to make social interaction successful, normal humans develop complex abilities in areas of social cognition, interpersonal skills and reasoning. A principal psychological construct that underpins social functioning is ‘Theory of Mind’ (ToM), which is the capacity to attribute independent mental states to others, and to explain and predict other people’s behaviour on the basis of their mental states. It is therefore a way of making sense of others’ behaviour. ToM is culturally invariant, and demonstrates increasing complexity through childhood development over the first ten years of life, with little individual variation and narrow time frames. The earliest stage of ToM is demonstrated in infants at about 18 months of age when joint attention and pro-declarative pointing develops. From the age of 18 months to two years children start to grasp the idea of pretend play and by two years have a firm understanding of the concept of desire. Between the ages of three and four they begin to understand that others may not know what they know and therefore may hold false beliefs (1st order ToM; Wimmer & Perner, 1983). At the age of six or seven, children can understand that someone can hold beliefs about another person’s beliefs (2nd order ToM; Perner & Wimmer, 1985). Between the ages of nine and eleven years children develop an understanding of a social ‘faux pas’ i.e. when something has been said that should not have been (Baron-Cohen, O’Riordan, Stone, Jones, & Plaisted, 1999). In order to understand that this type of social transgression has occurred one has to identify two mental states: first, that the person committing the ‘faux pas’ is unaware that they have said

something inappropriate, and second that the person hearing it might feel insulted or hurt. This requires both cognitive and affective mental state representation. Finally, the ability to recognise complex emotions and mental states in others from their facial expression, particularly solely from the eyes, has more recently been postulated as an advanced aspect of ToM (Lough, Gregory, & Hodges, 2001) and emerges around the time of adolescence.

Assessing ToM in People with FvFTD

ToM has a number of features that suggest it is a domain-specific (or modular) social cognitive ability and not just the result of general reasoning abilities applied to a social world (Brüne & Brüne-Cohrs, 2006). Firstly, ToM goes through the stereotypical developmental sequence, and secondly there is dissociation from other areas of psychological function. For example, people with Down's syndrome, where cognitive ability is impaired, perform in line with their mental age on ToM tasks; whereas people with (Autistic Spectrum Disorders) ASD are disproportionately negatively affected when carrying out ToM tasks compared with other aspects of intellectual function (Gregory *et al.*, 2001). The relationship of ToM to executive functioning is controversial. This is because ToM can place heavy demands on executive functioning, which has led some researchers to suggest that ToM impairment may be a result of executive deficits (Snowden *et al.*, 2003). However, wider research suggests that ToM is actually independent of overall neuropsychological status (Lough *et al.* 2006). Significantly, people with fvFTD generally perform well on tests of frontal lobe functioning indicating the relative insensitivity of these tests to accurately diagnose fvFTD. This leads to a good argument for the development of a ToM battery of tasks to aid early diagnosis of fvFTD.

RATIONALE AND METHODOLOGY FOR THE LITERATURE REVIEW

The aim of this review was to systematically evaluate research using ToM tests with people with fvFTD. If there is sound evidence that ToM is disturbed or impaired in this clinical group, then this may consequently assist in the development of effective neuropsychological testing procedures, potentially to aid in early diagnosis.

Searches were completed between June 2007 and September 2008. The databases that provided the most relevant literature were Psychinfo, Medline, and Embase. The two main areas of investigation were fvFTD and ToM. Combinations of the following key words were

used: FTD, Pick's disease, dementia of frontal type, frontotemporal lobar degeneration, ToM, social function / reasoning / conduct / cognition, executive function / deficit (See appendix 2 for details). The search parameters were set to find papers published in the last 10 years, since the Lund and Manchester criteria were developed (1998-2008; appendix 3). Research studies were included if they specifically evaluated the relationship between ToM and fvFTD and if they used standardised tests or criteria to measure or define these terms. No unpublished manuscripts or dissertation abstracts were included. One published poster presentation abstract was excluded because not enough detail was given about the study (Allegri *et al.*, 2006).

A total of 179 papers were identified of which nine fulfilled the criteria for inclusion in the review. The reference sections of these selected papers were hand-searched but no further papers were identified. The search did not identify any papers published before the year 2001. Notably, Kipps and Hodges (2006) had already published a conceptual paper on the relationship between fvFTD and ToM. In their paper they refer to the most recent studies in the field. They discuss the nature of defective ToM ('false beliefs', 'faux pas' etc.) and its relationship to the concept of empathy in people with fvFTD. They then consider how impairments in these abilities may influence or relate to semantic knowledge, social rules, and moral reasoning. They go on to explore the issue of executive function and its relationship to ToM, and then discuss the association of emotional processing deficits with ToM. They then take a clinic-anatomical perspective and contemplate the issues they discuss in terms of dysfunction in regional brain structures, specifically dysfunction in the orbitofrontal and medial prefrontal cortex. They conclude that, although complex, the study of ToM abilities in people with neurodegenerative diseases has been valuable in contributing to the understanding of the functional processes involved. Whilst their paper was not suitable for inclusion in this review, all the papers covered in it are appraised *a priori* here. This appraisal aims to provide a more systematic synthesis of the evidence.

Table 1: Overview of Papers: Design and analyses

No.	Authors	Year	Country	Design	Analyses
1.	Lough, S., Gregory, C., & Hodges, J.R.	2001	UK	Case study	Unspecified
2.	Gregory, C., Lough, S., Stone, V. Baron-Cohen, S., Hodges, J.R.	2001	UK	Cross-sectional between groups & intercorrelations	T-tests (fvFTD vs normal controls) for all ToM tests. Intercorrelations between ToM, executive tests and MMSE.
3.	Gregory, C., Lough, S., Stone, V., Erzinclioglu, S., Martin, L., Baron-Cohen, S., & Hodges, J.R.	2002	UK	Cross-sectional between groups & correlational	ANOVA (fvFTD, AD, normal controls); correlations between ToM and Neuropsychiatric inventory (NPI) tests, and degree of atrophy and ToM
4.	Lough, S. & Hodges, J. R.	2002	UK	Case study	Unspecified
5.	Snowden, J. S., Gibbons, Z. C., Blackshaw, A., Doubleday, E., Thompson, J., Craufurd, D., Foster, J. Happé, F., & Neary, D.	2003	UK	Cross-sectional correlational between groups & within groups comparisons	Repeated measures ANOVA for (fvFTD, HD and controls) performance on all 4 tests of social cognition (ToM). T-tests for error types. McNemar test χ^2 for chance level performance of 3 participants in MIE test. Correlations between social cognition and standard executive tests, and between fvFTD performance and distribution of frontal atrophy.

No.	Authors	Year	Country	Design	Analyses
6.	Lough, S., Kipps, C.M., Treise, C., Watson, P., Blair, J.R., & Hodges, J. R.	2006	UK	Cross-sectional between groups comparisons	One way ANOVA for group (fvFTD v Control) comparisons on executive tasks. ANCOVA for 2 group vs 2 cartoon types (ToM and Physical) & 2 group vs 2 story types (ToM and Physical) (executive scores as covariates). ANOVAS with moral task, emotions. ANCOVA with social situations. Wilcoxon signed ranks with IRI.
7.	Eslinger, P.J., Moore, P., Troiani, V., Antani, S., Cross, K., Kwok, S., & Grossman, M.	2007	USA	Cross-sectional between groups & correlational	ANOVA (fvFTD v APH, Total FTD v Controls). Regression analysis – correlations between all measures.
8.	Torralva, T., Kipps, C.M., Hodges, J.R., Clark, L., Bekinschtein, T., Roca, M., Calcagno, M.L., & Manes, F.	2007	Argentina	Cross-sectional between groups & correlational (within group?)	T-tests for most neuropsychological data (Mann Whitney <i>U</i> tests if not homogenous). MIE test: T-tests between MIE, fvFTD, Controls. False belief: Repeated measures ANOVA on correct hits v rejects x group, and also affective vs intentional x group. Spearman's <i>r</i> - correlating decision making, ToM, and executive functioning.
9.	Kosmidis, M. H., Aretouli, E., Bozikas, V.P., Giannakou, M., & Ioannidis, P.	2008	Greece	Cross-sectional between groups comparisons	Mann Whitney <i>U</i> tests on group means (control v fvFTD, and control v schizophrenia).

RESULTS

In the following sections each principal aspect of the nine identified studies is described and evaluated, and the main findings are summarised.

Study Aims

Lough *et al.* (2001) first developed the hypothesis that behavioural disturbance in fvFTD might be a reflection of the loss of ToM ability, and linked this to the literature based on the development of social behaviour in children. Then, in the studies by Gregory *et al.* (2001; 2002) that hypothesis was tested by examining the differences, first with healthy controls and then in comparison with people with Alzheimer's disease. They looked at performance on ToM tasks and executive tests, and assessed behavioural and neuropsychiatric dysfunction. These relationships were discussed further in a subsequent paper where they reported on just one participant from their study (Lough & Hodges, 2002). Next, the ability to interpret social situations was the focus of Snowden *et al.*'s (2003) paper, in which they hypothesised that people with fvFTD would exhibit a particularly marked impairment when this also involved attributing a mental state to another person. A wider perspective was then taken by Lough *et al.* (2006) who examined the relationship between ToM, emotional recognition and responsiveness, and abilities in empathising and moral reasoning; they predicted that people with fvFTD would show deficits in all these areas.

A model of social cognition was presented by Eslinger *et al.* (2007) who proposed that executive resources and social knowledge, through a social executor framework, guide interpersonal decision making. They stated that components of social knowledge (such as social perception, reasoning, and the understanding of consequences) were intimately linked with ToM processing. They included a measure of empathy, as they suspected that empathic sensitivity would effect processing of social information, and explored the model by examining how people with fvFTD resolve 'standardised' social dilemmas. In a similar vein, Torralva *et al.*'s (2007) premise for their research originated from the idea that the same underlying brain structures that are damaged in people with fvFTD, are responsible for ToM processing *and* affective decision making. They therefore expected that performance on the Iowa Gambling Task (IGT; Torralva *et al.*, 2007) would correlate with that on ToM tests, and

they wanted to assess the sensitivity of the IGT to highlight this relationship in people with fvFTD.

Finally, Kosmidis *et al.* (2008) were interested in the relationship between frontal and temporal impairments, and difficulties with social interactions, that are found in both people with fvFTD and schizophrenia. They compared performances on tasks that draw on ToM abilities and hoped that any differences in the patterns of deficit that emerged might reveal the point in the process of ToM in which a breakdown occurs.

Study Designs

Two papers were based on observational case studies of men with fvFTD (Lough *et al.*, 2001; Lough & Hodges, 2002). It is difficult to generalise from single case studies, however, they are an excellent way to document initial observations which can lead to the development of hypotheses, testable in larger studies and generalised to the wider fvFTD population. The case studies were published early in the history of this area of research.

The other seven studies were cross-sectional observational designs that compared differences between groups; either with healthy controls or with other clinical populations (e.g. Huntington's disease, Alzheimer's disease) or both. Unlike the others, the Lough *et al.* (2006) and Kosmidis *et al.* (2008) studies did not carry out any within-group statistical correlations; they solely conducted between-groups analyses. Of those that did investigate correlations, Snowden *et al.* (2003) and Torralva *et al.* (2007) reported that they performed within group correlations on for example: executive functioning and ToM abilities; or decision making ability, ToM tasks, and executive tests. The other papers did not specify that they used within-groups analyses (Gregory *et al.*, 2001; Gregory *et al.*, 2002; Eslinger *et al.*, 2007).

The results obtained from the cross-sectional designs offer more generalisable information, when compared with single cases, on account of the statistical evidence. However, the way in which the cross-sectional study is designed and conducted relative to the researchers' aims and hypotheses will have an effect on the validity of the conclusions that can be drawn. For example, cross-sectional studies often fail to 'control for' confounding factors that affect or even determine the relationship between the supposed cause and effect (e.g. in the current context: conducting tests over time, on different days, and with different researchers). Furthermore, correlational analyses look for relationships between variables

only (e.g. between ToM measures and executive functioning) and therefore cannot ascribe causality. The next section will deal more with these issues.

Table 2: Sample Characteristics: FvFTD, Controls, and Comparison groups

Study No.	1	2	3	4	5	6	7	8	9
Authors	Lough <i>et al.</i> (2001)	Gregory <i>et al.</i> (2001)	Gregory <i>et al.</i> (2002)	Lough & Hodges (2002)	Snowden <i>et al.</i> (2003)	Lough <i>et al.</i> (2006)	Eslinger <i>et al.</i> (2007)	Torralva <i>et al.</i> (2007)	Kosmidis <i>et al.</i> (2008)
fvtFD	n=1	n= 13	n= 19	n=1	n=13	n=18	n=12	n=20	n=9
Age (yrs)	47	44-67	44-67	57	Mean=60	47-74	Mean=66	Mean=67	Mean=65
Sex (M/F)	1 / 0	No info	16 / 3	1 / 0	9 / 4	16 / 2	No info	11 / 9	No info
Controls	n/a	n=13 age and education matched volunteers	n= 16 healthy	n/a	n= 18 healthy spouses of the patient groups	n= 13 matched healthy	n=17 age and education matched	n=10 age and education level matched healthy	n=10. age and education matched healthy
Age (yrs)	-	No info	52-76	-	Mean = 49	43-75	Mean=75	Mean=63	Mean=64
Sex (M/F)	-	No info	8 / 8	-	8 / 10	9 / 4	No info	4 / 6	No info
Comparis on group (No.)	-	-	n=12 Alzheime r's Disease	-	n=13 Huntington 's Disease	-	n=14 Progressive aphasia (APH)	-	n=28 Schizophreni a compared with 26 controls Mean= 36
Age range	-	-	52-79	-	Mean= 50	-	Mean= 71	-	
Sex (M/F)	-	-	6 / 6	-	5 / 8	-	No info	-	No info

Methods

In order to evaluate the research, attention needs to be drawn to the procedure followed by each study. The purpose of the methods section is to explain exactly what procedures were used to collect the data and, importantly, there should be sufficient detail to allow a precise replication of the study. There should also be adequate information regarding the sample, for example, the inclusion and exclusion criteria, the sampling procedure or recruitment method, the sample size, and a description of the important characteristics of that sample (e.g. age, sex). An assessment of methodology in relation to the selection and administration of tests is reported below (see 'Measures' section). Firstly, attention is drawn to the participants.

Participants (Table 2)

1. FvFTD groups: Inclusion criteria / fvFTD diagnosis

The Lund and Manchester criteria for diagnosing fvFTD were devised and published in 1998 in order to provide the foundation for research work (Neary *et al.*, 1998; appendix 3). Six studies specifically stated that they used these criteria (Gregory *et al.*, 2002; Snowden *et al.*, 2003; Lough *et al.*, 2006; Eslinger *et al.*, 2007; Torralva *et al.*, 2007; Kosmidis *et al.*, 2008). The exceptions were the two case studies (Lough *et al.*, 2001; Lough & Hodges, 2002) which did not quote the criteria specifically. The other study (Gregory *et al.*, 2001) was originally published as a poster presentation, and whilst they stated that their participants fulfilled 'consensus criteria' they did not specify which these were. In any case, all three of these studies originate from the Cambridge research group, which reports using the criteria in all their other fvFTD research.

The Cambridge group have also developed their own locally agreed criteria for screening and recruiting to their research trials. They require a clinical interview involving a detailed history and assessments by three independent academic and health professionals (professor of neurology, consultant psychiatrist, and consultant psychologist). Participants also undergo structural CT, MRI, and HMPAO/SPECT imaging, and although imaging is not necessary according to the consensus criteria, it can enable a more confident diagnosis of fvFTD and importantly rule out other aetiologies. In a similar fashion, the other four papers (Snowden *et al.*, 2003; Eslinger *et al.*, 2007; Torralva *et al.*, 2007; and Kosmidis *et al.*, 2008) also carry out all or

various combinations of neurological examinations, neuropsychological assessments, and structural imaging techniques. Thus all studies appear to have used rigorous diagnostic criteria.

2. FvFTD groups: sampling / recruitment

Not much detail was provided on how participants were recruited in any of the studies, but studies did usually report on where they recruited from. The Cambridge research group usually documented this information well. In their comparison studies, two papers (Gregory *et al.*, 2002; Lough *et al.*, 2006) stated that they had recruited from the early onset dementia clinic at Addenbrookes Hospital in Cambridge. The third comparison study, being a more concise poster report (Gregory *et al.*, 2001), omitted information about recruitment altogether as did the first case study (Lough *et al.*, 2001). The second case study in contrast (Lough & Hodges, 2002) recruited an inpatient. Of the remaining experimental designs, the final paper which originated from the UK, by Snowden *et al.* (2003), stated that they recruited from a Neurology Department Specialist Dementia Clinic in Manchester, and Torralva *et al.* (2007) from Argentina recruited at the Cognitive Neurology Division at Raúl Carrea Institute for Neurological Research in Buenos Aires. Neither the American (Eslinger *et al.*, 2007) nor the Greek researchers (Kosmidis *et al.*, 2008) gave details on their recruitment process.

It is important to consider the relationship between where participants were recruited, and where the testing was carried out, and how this might impact on the type of participants consenting to be in the study; whereas recruitment in an outpatient clinic might seem a more usual and user-friendly procedure, recruitment through a renowned academic research institute may bias the type of people who consent to take part. In addition, not everyone may be willing to travel to additional appointments.

3. FvFTD groups: Sample size

Aside from the case studies (Lough *et al.*, 2001; Lough & Hodges, 2002), the other seven studies recruited between nine and 20 fvFTD participants and compared them with between 10 and 18 controls. Small samples may affect the statistical power and are also more likely to introduce bias than larger samples. For instance, given a sample size of five, one deviant response in a group this small will represent a bias equivalent to 20% of the sample size; but in a group of 30 one deviant response only

represents just over 3% as a proportion. However, increasing the sample size can also mask poor methodology, in that if there are a large number of uncontrolled factors, the influences they exert may tend to be cancelled out. In these studies, where it is reasonable to expect the independent variable to exert a similar influence on all people, a sample size of 25 to 30 would be considered adequate. In fact, a small sample with good controls would be the best way of identifying hypothesised differences between groups. Furthermore, in studies such as the ones under review here the clinical populations are relatively small and so this is perhaps naturally reflected in the size of the groups.

4. FvFTD groups: Sample gender and age

Three studies did not report any information on the sex of their participants (Gregory *et al.*, 2001; Eslinger *et al.*, 2007; Kosmidis *et al.*, 2008). Of the studies that did report this information, there were more men in the fvFTD groups, in fact in Lough *et al.*'s (2006) study there were 16 males and only two females in their clinical group. In terms of age, three studies (Gregory *et al.*, 2001; Gregory *et al.*, 2002; Lough *et al.*, 2006) presented the age of their participants as a range in comparison to the other four studies that reported a mean (Snowden *et al.*, 2003; Eslinger *et al.*, 2007; Torralva *et al.*, 2007; Kosmidis *et al.*, 2008). The mean age of participants with fvFTD in Snowden *et al.*'s (2003) study appeared to be significantly higher than that for controls or the comparison group.

5. Control groups

Without adequate control groups the findings of a study may be suspect. The control and/or comparison group should be similar and matched on all the factors that are considered to be of importance, for example in these studies the paramount concerns appeared to be age, sex, and educational achievement. This is necessary in order to control for any extraneous factors which may cause confounding problems. In practice the sample for one group is usually selected using a standardised selection procedure, and then members of the second group are selected according to the matching criteria established. Also, to be representative the age range of both groups should reflect that of the population to which the results will be generalised.

In most of the studies the controls were generally referred to as 'healthy'. The Cambridge group clearly described their participants and stated that they all underwent full neuropsychological assessment. In two studies the exclusion criteria were made explicit and specified, for example Snowden *et al.* (2003) stated that controls should have no history of neurological disease, head injury, or alcohol abuse. Kosmidis *et al.* (2008) took this a little further and presented the following exclusion criteria: no psychiatric or neurological illness, no diagnosis of a developmental disorder or history of head injury with loss of consciousness, no alcohol or drug abuse during the six months prior to testing, and no physical illness that could affect cognitive performance.

The majority of studies stated that they had matched their controls on the criteria of age and education, but this was generally not elaborated upon. Nevertheless, while it appeared that most were well-matched, Gregory *et al.* (2002) reported statistically significant differences in age and education levels between their clinical and control groups and endeavoured to control for this by using ANCOVA in their analysis. In addition, Eslinger *et al.*'s (2007) controls were slightly older than their clinical group and unfortunately they did not acknowledge that this could potentially affect their results.

In terms of where the controls were recruited from, Torralva *et al.* (2007) stated that they were selected from the same geographical area as the patients, and Gregory *et al.* (2001) specified that volunteer controls were drawn from the Medical Research Council - Cognition and Brain Sciences Unit subject panel. Interestingly, Snowden *et al.* (2003) drew their control group from the pool of spouses of the participants in both their patient groups.

Finally, in reference to the gender composition in the control groups, two studies as already mentioned above completely neglected to report information about the sex of their participants (Eslinger *et al.*, 2007; Kosmidis *et al.*, 2008). Nonetheless, where information was made available the ratio of males to females were generally less marked in the control and comparison groups, than in the participants with fvFTD.

Procedure

It is important to consider the nature of testing overall. Some studies required that participants undertake many tests. This raises issues of how the researchers accounted for fatigue, order effects, and the continuity of test administrators from day-

to-day. Were follow-up sessions required and were these planned on consecutive days or over a period of weeks? What may have occurred during this time period to interfere with performance? Perhaps there were other factors specifically related to the environment that may have affected the result obtained. Also, with this client group the nature of symptoms may mean that they refuse to comply with the testing process, how was this overcome? Typically people with fvFTD have problems with attention and concentration; did researchers report an attrition rate or any drop out statistics? Furthermore, how ethical is it for researchers to subject people with fvFTD to extensive psychological testing? It also needs to be noted that the validity of findings where the participants or experimenters know the aim of the study are always doubtful, as the experimenters will tend to find what they are looking for. Generally, studies did not report this kind of information, or refer to it in their discussion, but all would be important considerations to take into account when designing and conducting a research project with this clinical population.

Measures

1. ToM tasks

There are a number of tasks commonly used for the assessment of the development of ToM in children, specifically the ‘first order false belief test’ (Wimmer & Perner, 1983), the ‘second order False Belief Test’ (Wimmer & Perner, 1985), the ‘Faux Pas test’ (Stone, Baron-Cohen, & Knight, 1998) and the ‘Mind-in-the-Eyes Test’ (Baron-Cohen, Jolliffe, Mortimer, & Robertson, 1997). These are all used in the four earliest studies reviewed here (Table 3), the majority of which are by the Cambridge group. The tests have the advantage of being well known, of being straight forward to understand conceptually, and their names are descriptive enough for the reader to appreciate which aspect of ToM each one tests. They test progressively more complex aspects of ToM and so give a thorough overview of participants’ ToM ability or deficits. These early studies clearly established the presence of ToM deficits in fvFTD. A fifth study (Torralva *et al.* 2007) used only the ‘Faux Pas test’ and the ‘Mind-in-the-eyes test’ in their comparison of ToM and affective decision-making.

Only one of the other studies uses a set of several tests (Snowden *et al.* 2003). The tests chosen appear to assess 1st and 2nd order false beliefs and the sophisticated ToM level of reading intention from the eyes (Happé, Brownell, & Winner, 1999). It is

not entirely clear from the descriptions whether understanding of social unacceptability (*faux pas*) was tested through their assessments. In addition, all items in their 1st and 2nd order tests depend on use of humour as well as appreciation of others' beliefs thus giving a less general assessment of basic ToM. Overall, therefore Snowden *et al.*'s choice of measures does not seem as conceptually clear as those by the Cambridge group.

The other three studies use one or two ToM assessments each. The purpose of Lough *et al.*'s (2006) paper is specifically to broaden understanding, by extending exploration to advanced aspects of ToM not hitherto assessed in fvFTD, i.e. the aspects of moral reasoning and recognising violations of socially unacceptable behaviour. Their choice of measures from tests developed for schizophrenia (Corcoran, Cahill, & Frith, 1997) and for a wide range of children and adults (Happé, 1994) is well justified. In contrast, Kosmidis *et al.* (2008) use a novel means of assessing ToM which uses video tapes of actors rather than pictures to assess 2nd order inferences (McDonald, Flanagan, Rollins, & Kinch, 2003). This is a restricted level of ToM but appears to be a sound method of assessment. Eslinger *et al.* (2007), who provide the only American study, also assess only 1st and 2nd order beliefs but with very clearly described assessments (Winner, Brownell, Happe, Blum, & Pincus, 1998).

Overall, given the complex nature of ToM, it is important that studies report which aspects of ToM are being tested. This is better addressed in some studies than others. The use of actors in a quasi real-life situation would appear to have some possible advantages due to its face validity, but such situations may also introduce some confounding aspects such as paralinguistic clues whereas cartoons do not introduce such variables. Whether cartoons or actors are used, it seems more thorough to use a variety of concepts i.e. humour, paradox, sarcasm and deception, rather than rely on only one of these. The studies by the Cambridge group appear to be of consistently high quality in these regards.

Table 3: ToM Measures

ToM Measure	Study Ref.	1. Lough <i>et al.</i> (2001)	2. Gregory <i>et al.</i> (2001)	3. Gregory <i>et al.</i> (2002)	4. Lough & Hodges (2002)	5. Snowden <i>et al.</i> (2003)	6. Lough <i>et al.</i> (2006)	7. Eslinger <i>et al.</i> (2007)	8. Torralva <i>et al.</i> (2007)	9. Kosmidis <i>et al.</i> (2008)
1st Order False Belief	Wimmer & Perner (1983)	✓	✓	✓	✓					
2 nd Order False Belief	Wimmer & Perner (1983)	✓	✓	✓	✓					
Faux Pas	Stone <i>et al.</i> (1998)	✓	✓	✓	✓				✓	
Mind-in-the-Eyes	Baron-Cohen <i>et al.</i> (1997)	✓	✓	✓	✓	✓			✓	
Single Cartoon Abstraction	Happé <i>et al.</i> (1999)					✓				
Cartoon Pairs	Happé <i>et al.</i> (1999)					✓				
Story Comprehension	Happé <i>et al.</i> (1999)					✓				
Cartoons	Corcoran <i>et al.</i> (1997)						✓			
Stories	Happé (1994)						✓			
False Belief Vignettes	Winner <i>et al.</i> (1998)							✓		
Perception of Social Inference Test	McDonald <i>et al.</i> (2003)									✓

2. Other measurements

A number of measures of other aspects of cognitive functioning were used in the studies (table 4). These included well known and standardised neuropsychological tests, with reportedly good reliability and validity. The four earlier studies, carried out by the Cambridge group, include a fully comprehensive and extensive test battery designed to measure the domains of executive, frontal, and semantic functioning, and memory. They included tests of general cognitive functioning, estimates of pre-morbid IQ, and visuospatial functioning, and they also administered the Neuropsychiatric Inventory (NPI; Cummings *et al.*, 1994) to carers. One study (Gregory *et al.*, 2001) was a published poster presentation and did not list all measures used, but did report some test results.

Of the five more recent studies, only one reported administering comprehensive neuropsychological testing. Torralva *et al.* (2007) used a general cognitive test battery, extending this to include additional measures of executive and frontal functioning. Snowden *et al.* (2003) only reported using a general measure (MMSE) of functioning with the addition of two assessments of executive and frontal functioning (WCST & FAS verbal fluency). This may be because in their study they were concentrating on comparing the similarities and differences across their groups, between people with fvFTD and Huntington's disease, rather than making links between cognitive domains i.e. ToM and executive functioning.

Lough *et al.* (2006) were interested in the relationships between the different aspects of social functioning. To this end they employed tests that assess social and moral reasoning in addition to measures that tap the processing of emotions and recognition of empathy. They used the Hayling and Brixton test to measure executive and frontal functioning. In a similar vein, Eslinger *et al.* (2007) was also interested in social functioning and employed a test designed to assess 'ability to resolve social dilemmas' in addition to rating empathy in the same individuals. Unfortunately Eslinger *et al.* did not report on general level of cognitive ability in their sample. This was also true in the most recent paper by Kosmidis *et al.* (2008) who relied on the Lund and Manchester criteria to explain neuropsychological status. In fact they claim that all their fvFTD participants were in the early stages of the disease but do not offer an explanation as to how this was assessed or quantified. This may be a weakness in their study.

Overall, the studies used a variety of measures in their research. The earlier ones, specifically from the Cambridge group, administered a full test battery of neuropsychological

functioning in order to compare and contrast abilities in neuropsychological functioning with ToM ability. Later studies rely on the Lund and Manchester Criteria and CT/ MRI HMPAO/SPECT imaging to categorise their fvFTD participants. This is probably because later studies have broadened their exploration to include investigations of areas suspected to be associated with and affected by ToM abilities, for example; perceiving sarcasm, resolving social dilemmas, moral reasoning, and empathic ability.

Table 4: Other measures used

Domain	Study	1. Lough <i>et al.</i> (2001)	2. Gregory <i>et al.</i> (2001)	3. Gregory <i>et al.</i> (2002)	4. Lough & Hodges (2002)	5. Snowden <i>et al.</i> (2003)	6. Lough <i>et al.</i> (2006)	7. Eslinger <i>et al.</i> (2007)	8. Torvalva <i>et al.</i> (2007)	9. Kosmidis <i>et al.</i> (2008)
	Measures									
Neuropsychiatric functioning	NPI (Neuro-psychiatric Inventory)	✓	✓	✓	✓					
General Cognitive Functioning	MMSE (Mini Mental State evaluation)	✓	✓	✓	✓	✓		✓	✓	
	ACE (Addenbrookes Cognitive Examination)			✓	✓				✓	
Pre-morbid IQ (estimate);	NART (National Adult Reading Test)	✓		✓	✓		✓			
	WAT-BA (Word Accentuation Test - Buenos Aries)								✓	
IQ	WAIS-R (full) (Wechsler Adult Intelligence Scale)	✓								
Visuo-spatial functioning	REY figure	✓	✓	✓						
	VOSP (Visual Object and Space Perception) Battery		✓	✓	✓					

Table 4: Other measures used (continued)

Domain	Study	1. Lough <i>et al.</i> (2001)	2. Gregory <i>et al.</i> (2001)	3. Gregory <i>et al.</i> (2002)	4. Lough & Hodges (2002)	5. Snowden <i>et al.</i> (2003)	6. Lough <i>et al.</i> (2006)	7. Eslinger <i>et al.</i> (2007)	8. Torvalva <i>et al.</i> (2007)	9. Kosmidis <i>et al.</i> (2008)
	Measures									
Memory Tests	WAIS: Digit Span									
	a) forwards (attention / concentration)			✓	✓				✓	
	b) backwards (executive function)			✓	✓				✓	
	WMS-R (full) (Wechsler Memory Scale – Revised)	✓	✓	✓						
	WMS-R (story recall only)				✓				✓	
	REY figure	✓	✓	✓						
	WRMT (Warrington Recognition Memory Test)				✓					

Table 4: Other measures used (continued)

Domain	Study	1. Lough <i>et al.</i> (2001)	2. Gregory <i>et al.</i> (2001)	3. Gregory <i>et al.</i> (2002)	4. Lough & Hodges (2002)	5. Snowden <i>et al.</i> (2003)	6. Lough <i>et al.</i> (2006)	7. Eslinger <i>et al.</i> (2007)	8. Torvalva <i>et al.</i> (2007)	9. Kosmidis <i>et al.</i> (2008)
	Measures									
Semantic function	Pyramids & Palm Trees	✓		✓	✓				✓	
	GWST (Graded Word Synonyms Test)									
	Concrete and Abstract Word Synonyms Test		✓		✓					
	GNT(Graded Naming Test)		✓		✓					
	BNT (Boston Naming Test)								✓	
	The Token Test								✓	

Table 4: Other measures used (continued)

Domain	Study	1. Lough <i>et al.</i> (2001)	2. Gregory <i>et al.</i> (2001)	3. Gregory <i>et al.</i> (2002)	4. Lough & Hodges (2002)	5. Snowden <i>et al.</i> (2003)	6. Lough <i>et al.</i> (2006)	7. Eslinger <i>et al.</i> (2007)	8. Torvalva <i>et al.</i> (2007)	9. Kosmidis <i>et al.</i> (2008)
	Measures									
Executive & frontal function	WCST (Wisconsin Card Sorting Test)	✓	✓	✓	✓	✓			✓	
	F.A.S. (verbal fluency test)	✓	✓	✓	✓	✓			✓	
	BADS (Behavioural Assessment of the Dysexecutive Syndrome)	✓								
	RCPM (Ravens Coloured Progressive Matrices)				✓				✓	
	Hayling and Brixton Tests						✓			
	VVT (Visual Verbal Test; Abstraction & Cognitive Flexibility)							✓		

Table 4: Other measures used (continued)

Domain	Study	1. Lough <i>et al.</i> (2001)	2. Gregory <i>et al.</i> (2001)	3. Gregory <i>et al.</i> (2002)	4. Lough & Hodges (2002)	5. Snowden <i>et al.</i> (2003)	6. Lough <i>et al.</i> (2006)	7. Eslinger <i>et al.</i> (2007)	8. Torvalva <i>et al.</i> (2007)	9. Kosmidis <i>et al.</i> (2008)
	Measures									
Executive & frontal function (continued)	Trail making (Pt A: attention / concentration)								✓	
	Trail making (Pt B: executive function)								✓	
	WAIS (letters and numbers ordering)								✓	
	FAB (Frontal Assessment Battery)								✓	

Table 4: Other measures used (continued)

Domain	Study	1. Lough <i>et al.</i> (2001)	2. Gregory <i>et al.</i> (2001)	3. Gregory <i>et al.</i> (2002)	4. Lough & Hodges (2002)	5. Snowden <i>et al.</i> (2003)	6. Lough <i>et al.</i> (2006)	7. Eslinger <i>et al.</i> (2007)	8. Torvalva <i>et al.</i> (2007)	9. Kosmidis <i>et al.</i> (2008)
	Measures									
Miscellaneous	Social reasoning						✓			
	i) Moral-emotional distinctions test						✓			
	ii) Social situations									
	Facial emotion expression processing task						✓			
	IRI (Interpersonal Reactivity Index; test of empathy)						✓			
	Cartoon Predictions (resolving social dilemmas)							✓		
	Empathy ratings (3 questions)							✓		
	IGT (Iowa Gambling Test)								✓	

Analyses

The two case studies (Lough *et al.*, 2001; Lough & Hodges, 2002) offered an in-depth examination. They reported performance on ToM measures, neuropsychological testing and provided detailed histories. They then commented on the relationship between these findings and the pattern of atrophy observed on neuro-imaging. The study by Lough & Hodges (2002) was in fact a more detailed description of a patient from a previous study (Gregory *et al.*, 2002). It provided neuropsychological data gathered over a three year period, which indicated the cognitive decline associated with this condition. Unfortunately, they did not report results from ToM tests carried out at similar time points, which may have been a useful supplement.

Gregory *et al.* (2001) used between groups t-tests to show that the difference in performance on ToM tests between fvFTD and controls is incontestable. However, the controls performed at the ceiling on two of the four ToM tests and near the ceiling on the third measure, which means that there is no variation in their scores at all. The use of a parametric comparison statistic was possibly therefore inappropriate. Inter-test correlations found that ToM test scores were correlated and that ToM performance was also correlated with MMSE but not with executive functioning. Unfortunately it is not clear whether this was done with pooled data or with data from the fvFTD group only, probably because this is a summary of a poster presentation which lacks detail. In a subsequent paper by Gregory *et al.* (2002) a three group design (fvFTD vs. AD vs. healthy controls) was analysed with ANOVA. They reported fulfilment of assumptions for use of parametric tests and employed an element of covariance to account for differences between groups in education and age. In this paper the description and choice of statistical methods is full and considered, though again, the complete lack of variance in the control group would appear to violate assumptions for parametric tests. They also carry out correlations between various tests using parametric or non-parametric tests as appropriate.

Snowden *et al.*'s (2003) three groups design (fvFTD vs Huntington's disease vs and healthy controls) used repeated measures ANOVA to look at differences between the groups across the four measurements of ToM. Correlations were then carried out between the total performance on all social cognition tasks with the performance on executive tests. The results section, although organised and well-described, is lengthy, and the ToM tests used have a complicated scoring system which may inhibit a reader's understanding. However, this was offset somewhat by the inclusion of some graphed data.

Lough *et al.*'s (2006) study used a one-way ANOVA to compare groups on the executive tests (fvFTD vs healthy controls x Hayling & Brixton tests), which indicated that people with fvFTD performed significantly worse than controls. They therefore needed to use ANCOVA, with the Hayling or Brixton tests as a covariate, to compare the groups on ToM tests and their social situations test. However, in order to compare the groups on the moral / conventional distinctions test and the emotional recognition test they were able to use 2x2 ANOVA because the same interaction was not indicated. They analysed the IRI data with Wilcoxon signed ranks as it was nonparametric, and overall they described their procedure in a logical and clear order. In contrast, Eslinger *et al.* (2007) clearly summarised participant characteristics and test scores in a table and then reported statistical data and probabilities in the results section. However they did not provide much detail on their analytic procedure so it can only be assumed that the data was parametric and that an ANOVA was used. In their results Eslinger *et al.* also reported on a regression analysis that looked at the correlations between the VVT, cartoon predictions, and ToM tests.

Torralva *et al.* (2007) used t-tests to compare differences between groups and the MiE test, and then repeated measures ANOVA to assess group differences in response between 'affective' and 'intentional' components of the False Belief test. Some of their samples, however, were not of equal variance and in fact the fvFTD sample was twice the size of the controls. Furthermore, homogeneity of variance could not be achieved even after transforming the data so they chose to use a non-parametric test (Mann Whitney *U*) to compare some samples. As in previous studies they also conducted Spearman's *r* correlational analyses to assess the relationship between fvFTD, decision making, ToM, and executive functioning, but it was not clear if they correlated the data for the controls in addition to people with fvFTD or not. Furthermore, in this study an extensive array of comparisons seemed to have been carried out which may be cause for concern. The Mann Whitney *U* test was also utilised by Kosmidis *et al.* (2008) to look at differences between their groups but they gave no explanation as to why they chose it. Perhaps if the conditions were met for parametric tests then these would have provided a more credible conclusion.

To summarise, the papers that were clearest to follow were the ones that openly explained their approach to analysis and the Cambridge group had the most effective method of reporting this in their studies. It involved the inclusion of a small section, usually entitled 'statistical analysis', before coverage of the results. This section summarised the nature of the

data (i.e. parametric or non-parametric) and then indicated which tests would be used on which data. Correspondingly, this meant that they provided the reader with an overview of the procedure they used to address their aims and hypotheses, and also prepared the reader for the layout of the results section; this way of reporting the analyses and results seemed logical and reader-friendly. Moreover, when this information was made available, the reader was in a better position to evaluate the choice of statistical test, the results obtained, and ultimately the conclusions drawn from them. Without a clear understanding of this process the reader may be forced to make assumptions which may or may not be factual, and subsequently this may limit the credence of the research.

The sections above have considered the quality of the research on ToM performance in fvFTD. Whilst there are a number of different designs used, each with advantages and disadvantages, overall the quality of the nine studies reviewed is high. They use appropriate designs, sound diagnostic criteria, good measures and generally employ appropriate analyses. More robust findings were evident from research which comprehensively measured neuropsychological functioning and used established and well known ToM tests. Overall, all studies are of good enough quality for their results to be treated as valid.

DISCUSSION

In this final section, the main findings from the studies are summarised, limitations of the review are acknowledged, and suggestions are made for future research and for putting the implications into clinical practice.

Main Findings

Overall, the evidence strongly indicates that ToM is impaired in people with fvFTD. However the simpler 1st and 2nd order ToM tests could be passed in milder cases (Gregory *et al.*, 2001). Nevertheless, the majority of people with fvFTD failed on the more complex social ‘faux pas’ task and had difficulty with the MiE task regardless of the stage of the disease. Gregory *et al.* (2002) again found that the more complex ToM abilities were particularly sensitive to ToM impairment.

Lough & Hodges (2002) found that people with fvFTD achieved near normal scores on frontal executive functioning but showed marked deficits on ToM tests. This suggests a

dissociation between traditional executive function and social cognition. They suggest that it is the deficits in ToM that may therefore underlie the difficulties in social conduct and behavioural disturbance which characterise people with fvFTD rather than executive functioning per se. In fact Gregory *et al.* (2002) reported a significant difference between the psychiatric symptoms displayed by people with AD and those with fvFTD and they related this to the deficits evidenced by people with fvFTD in several aspects of ToM.

In contrast, Snowden *et al.*'s (2003) study found that performance was poor on all tasks administered not just those with a ToM component. Hence they suggested that their findings indicated impairments in ToM as a consequence of impaired executive function. They argue that general executive deficits will have an inevitable impact on performance on ToM tasks and they acknowledge that this may mask more specific deficits in ToM.

Lough *et al.* (2006) reported that knowledge of social rules was generally intact but moral reasoning, emotion processing, and empathy was defective. Interestingly they reported that emotion recognition was globally impaired but particularly for the emotions of 'anger' and 'disgust', which they claim may explain why people with fvFTD find it difficult identifying social violations. They also found that executive functions were impaired but that ToM ability was independent of this. They concluded that executive functioning acts supportively as a domain general skill in ToM but does not explain all aspects of abnormal ToM processing.

The people with fvFTD in Eslinger *et al.*'s (2007) study exhibited impairments in their judgements of social dilemmas, in addition to deficits in ToM, self-awareness of empathy, and cognitive flexibility. Furthermore, there was a strong correlation between these abilities which led Eslinger *et al.* to conclude that the deficits seen in people with fvFTD in resolving social dilemmas are related to depleted executive resources and social knowledge.

People with fvFTD were impaired in both ToM tasks and on the IGT, in Torralva *et al.*'s (2007) study; however there was no significant association found between them. Torralva *et al.* hypothesise that similar prefrontal circuitry is probably involved but that the cognitive domains may actually be independent. They go on to state that the IGT seems particularly sensitive to the cognitive dysfunction evidenced in early fvFTD, and conclude that the deficits in decision making (IGT) and ToM that although distinct have additive effects on the development of social behaviour in people with fvFTD. Finally, in Kosmidis *et al.*'s (2008) study people with fvFTD generally performed poorly on the 'comprehension of

sarcastic remarks' subtest and in the 'lie' condition when they had to rely on paralinguistic clues indicating sarcasm or lies. When they were given additional verbal clues their performance improved.

In summary, these studies have all indicated that ToM is abnormal in fvFTD and that, furthermore, some aspects of ToM processing are more disrupted than others. For example ToM tasks that require 'affective' processing are more impaired than those that require more 'cognitive' ToM processing. Moreover, while executive functions seem to play a supporting role in ToM performance, to some degree they appear to dissociate.

Clinical Implications

The research looking at the relationship between performance on ToM tasks and FvFTD has implicated impairments or deficits in ToM early on in fvFTD, independent of executive functioning. It is therefore recommended that neuropsychological assessments are developed, specifically in early onset dementia clinics, to take this into account in order to aid early detection. Also, given that diagnostic problems occur early on in this disease, it may be appropriate for accurate and detailed information on this condition to be more widely available through primary care health services, in this way helping to improve the care pathway.

Limitations of this Review

Attention is drawn to some of the limitations of this review. Firstly, Kipps and Hodges (2006) have already published a conceptual review of all the literature on the relationship between fvFTD and ToM published up to 2006, which included all their own papers and the research by Torralva *et al.* (2007; accessed online in 2006). The only new piece of research was from Kosmidis *et al.* (2008). Therefore this review does not advance theoretical knowledge but instead focuses on the systematic appraisal of the methods used and the quality of the published reports. This does however highlight the fact that there is still limited research in this area. Most of the research in the field has been carried out in the UK by the Cambridge group, and it may be important for other researchers from different locations and cultural backgrounds to explore this area and attempt to replicate findings.

Relatively little emphasis was placed on anatomical / morphological concerns, in this review. Instead interest lay in the psychological effects evidenced in people with fvFTD

produced by changes in ToM and social functioning. However, recognition must be given to the studies reviewed here that considered the link between frontal lobe degeneration (indicated in structural imaging) on both ToM and fvFTD and the possibility of related brain circuits underlying and driving social functioning.

Conclusion and Future Directions

The studies in this review have provided a valuable insight into the contribution that ToM may have to understanding the impaired social functioning seen in this clinical group. Furthermore, it has been shown in imaging studies that the degenerative process in fvFTD affects other structures that have been shown to be intimately associated with ToM processing in the normal brain. For example, significant deficits in emotional processing are seen in fvFTD in addition to impairment in ToM ability. However, these deficits do not completely explain why empathy is affected so fundamentally in these people, or explain how an individual's knowledge of social rules can remain intact while the processing of rule violations, and the ability to be flexible about the ensuing consequences, can be impaired. Further work needs to be carried out in this area looking more specifically at the nature of these deficits, and at identifying tests sensitive to the early detection of ToM in early stages of degeneration while other cognitive functions are still preserved. More information is also needed on the different aspects or domains of social cognition and on the contribution that these make to the understanding of ToM and hence the behavioural changes seen in people with fvFTD.

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Research Paper

The Family Experience of Frontal-variant Frontotemporal Dementia: A Qualitative Study

ABSTRACT

Purpose: This research examines the experiences of people who have a family member with a diagnosis of frontal-variant frontotemporal dementia (fvFTD). The research aimed to address two broad questions: How does the development of fvFTD in a working age person affect the family experience of living with that person, and how might mental health services respond to the needs of those family members?

Methods: Semi-structured interviews were conducted with six participants and the resulting transcripts were subjected to Interpretative Phenomenological Analysis (IPA).

Results: Four main themes emerged from the data; ‘the opening of the eyes’ and ‘the double-edged sword’ were subsumed under the super-ordinate theme ‘emergence and realisation’ and related to the experience of becoming aware of fvFTD and entering the medical and social care system. The themes of ‘the adaptation’, and ‘the maintenance’, grouped together under the super-ordinate theme of ‘life adjustments and coping’, reflected the experience of coming to terms with a changing relationship, becoming a carer, and surviving it.

Conclusion: Family caregivers of people with fvFTD have to contend with specific behavioural challenges and personality changes associated with the condition. Knowledge about fvFTD is lacking in both carers and professionals alike causing uncertainty and long periods before diagnosis, which adds to the burden of care for these people. Services need to be developed to cater for specific individual needs and awareness needs to be raised in all health care services.

Key words: Burden of care, family caregiver, frontal-variant frontotemporal dementia, Interpretative Phenomenological Analysis, qualitative research

INTRODUCTION

The term dementia refers to a progressive decline in cognitive function caused by damage or disease in the brain, which is beyond what might be expected from normal aging. Frontotemporal dementia (FTD) is the fourth most common type of dementia affecting older people (Sjögren & Anderson 2006) and accounts for approximately 20% of cases (Snowden, Neary, & Mann, 2002; Graham & Hodges, 2005). It is also the second most common form of early onset dementia (occurring before the age of 65) after Alzheimer's disease. Notably, recent studies have found FTD to be a more common cause of early onset dementia than was previously recognised (Ratnavalli, Brayne, Dawson, & Hodges, 2002; Rosso, Katt, & Baks, 2003).

FTD can be differentiated from other dementias on the primacy of behavioural and personality changes and the progression of language dysfunction in the absence of memory impairments. Classification has been confusing due to the use of different terms but in 1998 consensus criteria were published. The Lund and Manchester Groups (1994) had previously published a list of core diagnostic features and Neary *et al.* (1998) used this as a foundation to divide dementia of the frontal lobes into three clinical sub-types (FTD, semantic dementia, and progressive non-fluent aphasia) based on core and supportive diagnostic features (Neary *et al.*, 1998; see appendix 3). It is these consensus criteria which this study subscribes to. There are other classification systems but regardless of the system subscribed to, the progression and prognostic timeframe of FTD is the same.

In people with FTD, degeneration is located primarily in the frontal lobes and consequently the term frontal-variant FTD (fvFTD) has commonly been used to label their dementia. They present with changes in personality and behavioural disturbances; interpersonal difficulties characterised by a lack of empathy or concern for others; disinhibition or other socially inappropriate behaviours; and a general lack of insight and apathy. Due to the prominence of behavioural symptoms and changes, fvFTD is also often termed behavioural variant FTD (bvFTD). In addition, the progressive social impairments together with some executive deficits which are also a prominent clinical feature of fvFTD have led to the use of the term 'social and executive disorder' (SOC/EXEC; Eslinger *et al.*, 2007). Some researchers have labelled these behavioural problems as 'sociopathic' (Mendez,

Chen, Shapira, & Miller, 2005), whereas others report aggressive, socially disruptive, and antisocial behaviour (Miller, Darby, Benson, Cummings, & Miller, 1997).

Onset of fvFTD is insidious and according to clinical studies occurs most often in the age range of 45-65 (Sjögren & Anderson, 2006; Snowden, Neary, & Mann, 2007), although it can present before the age of 30 and in the elderly (The Association for Frontotemporal Dementias [AFTD], 2006). Dementia health services are currently set up to respond predominantly to memory-related problems and 'Memory Clinic' services are therefore oriented and equipped to assess and diagnose dementias where memory problems typically occur earlier, such as in Alzheimer's disease (AD) and vascular dementia (VaD; The Alzheimer's Society, 2006). Due to the predominance of over 65s with AD and VaD these are also therefore primarily older peoples' services. As already indicated, people with fvFTD eventually do develop memory difficulties but typically much later on, so initially fvFTD is often misdiagnosed as an affective disorder (such as depression), psychosis, or alcohol abuse (Sjögren & Anderson 2006). After the eventual development of memory difficulties, people with FvFTD also experience more global impairments with deterioration in intellect and language skills. Specific symptoms may vary from patient to patient but progression is inevitably downhill, ranging from less than two years in some people to more than 10 years in others (AFTD, 2006). It follows therefore that a person living with fvFTD experiences and exhibits a unique and different symptom pattern from those with other dementia types. This would, as a result, logically require qualitatively different interventions or support structures.

In terms of care for people with dementia, the majority are looked after at home by family caregivers, particularly spouses (Lewis, 1998). This places a heavy demand on them, in terms of their emotional and physical wellbeing, due to the immense strain and distress resulting from trying to manage the aberrant behaviours and other symptoms related to the dementia (Harvath, 1994). Moreover, there are also associated disruptions in family, work, and social relationships to contend with (Donaldson, Tarrier, & Burns, 1997). This phenomenon is referred to as 'burden of care' and there is a complex relationship between the aberrant behaviour and the negative consequences for carers. Some carers report higher levels of strain than others and research has identified various intrapersonal variables which may mediate the relationship between the stressful aspects and the negative consequences. For example, Tarrier *et al.* (2002) suggested a relationship between carers' perceptions and attributions of behaviour and the degree of their distress. They reported that 'critical'

caregivers were most likely to construe negative behaviours and situations as within the control of care recipients. They indicated that caregiver characteristics were as important as, if not more, than care receiver characteristics in shaping their emotional responses and their explanations of the symptoms and behaviours. More recently, Campbell *et al.* (2008) have reported that it is not the 'objective' load from the patient but the 'subjective' interpretation by the carer and the adopted coping style that subsequently determines the burden levels. So essentially research has shown that the variation experienced by carers can often be explained by differences in how they perceive, interpret and then manage the problem behaviour.

There is relatively little research about fvFTD that does not focus on clinicopathological assessments or genetic investigations. However, a small study in Japan of two people with fvFTD (Kumamoto, Arai, Hashimoto, Ikeda, Mizuno, & Washio, 2004) looked at the problems that family caregivers encountered while caring for their relatives at home. They reported that the very specific behavioural symptoms typical to people with fvFTD create major problems and a heavy burden for family carers. These symptoms include inappropriate social behaviours like shouting, swearing, and aggressive behaviour, making inappropriate comments in public, taking jokes too far, and exhibiting sexually inappropriate behaviour. The authors concluded that more resources need to be allocated to the specific needs of people with fvFTD and their families.

Rationale

The rationale for this particular study is based on the fact that more information is needed about the impact of fvFTD on partners, spouses and other family members of those with the diagnosis. At present services are designed to cope with memory problems. Information from this study may assist services to develop interventions that specifically respond to the social functioning and behavioural issues related to fvFTD, which are qualitatively different from dementia of other types.

This research takes an in-depth look at the experiences of family members in caring for someone with fvFTD. It utilises a qualitative approach which acknowledges the researcher's personal perspective and limits the constraints placed on the participants' responses. The aim is to uncover a broad and rich array of information which will add to the knowledge base.

METHODOLOGY

Study design and methodology

The research took a qualitative approach with semi-structured interviews being taped and transcribed verbatim, and the data analysed according to Interpretative Phenomenological Analytic procedure (IPA; Smith, Jarman, and Osborn, 1999; Willig, 2001).

Philosophically underpinning IPA is the view that meaning essentially occurs through understanding subjective experiences (or phenomena), and not through the accumulation of supposedly objective ‘facts’. It follows therefore that in order to understand human experience it is necessary to explore the nature of that experience as closely as possible (McLeod, 2001). IPA consequently explores individuals’ views of the topic under investigation and the meanings that they ascribe; essentially trying to understand how they make sense of their experiences. These meanings together with the researcher’s subsequent interpretations of them can be regarded as ‘social constructions’ rather than objective truths. So it follows that there is no ‘right’ interpretation, as each individual will have a different view or ascribe different meanings. IPA considers that many interpretations are possible. It takes an idiographic approach and requires the researcher to analyse participants’ transcripts one by one. The researcher develops insight into the meaning of the experience or event as a direct result of full immersion or engagement with the text. The insights and interpretations are integrated in the end stage of the analytic procedure.

Participants

For inclusion participants had to be a relative of a person who had received a diagnosis of fvFTD (according to the Lund-Manchester criteria; The Lund and Manchester Groups, 1994) from a working age dementia service. They also needed to speak and understand English, as the methodology required direct communication and an analysis of that communication. The presence of a translator and the translated material would therefore have impacted on the validity of the analysis. In all, six family members of people with fvFTD were recruited (Table 1.).

Table 1: Summary information on participants

participant	pseudonym	sex	relationship	age
1	Mr Smith	m	husband	67
2	Mr Jones	m	husband	58
3	Mrs White	f	wife	59
4	Mr Brown	m	husband	58
5	Miss Green	f	daughter	23
6	Mr Mills	m	brother	39

Procedure

The researcher attended team meetings with two specialist working age dementia services in the UK in order to introduce the project and enlist interest and support. Professionals in those services then identified suitable participants from their outpatient lists. They raised the idea of taking part in the research with their clients' partners, to avoid 'cold calling' by the researcher and to maintain confidentiality. If they were interested then they were given an introductory letter and information sheet (appendix 8). After a minimum of 24 hours had elapsed the researcher contacted them by telephone to answer any questions, gain consent and arrange to carry out the interview.

The Researcher met each participant on a one-to-one basis to conduct an in-depth semi-structured interview. One participant, however, chose to have their relative present, who was the person diagnosed with fvFTD. Interviews were arranged to occur in a place that was convenient for participants so for most interviews this involved meeting in the participant's own home, except for one who was interviewed at the care facility in which his relative was resident. A signed written consent form was obtained prior to the start of the interview (appendix 9). The interview schedule was initially developed from, and informed by, the results of an unpublished small scale study carried out locally and entitled: 'A qualitative study using focus groups to establish services required by carers of people with dementia-related changes to their social functioning'. To begin the interview process, brief demographic data were obtained which initially also helped to establish rapport. The interview schedule then included questions designed to tap into various areas i.e. health and history, diagnosis, living arrangements, day-to-day life, and the effects or consequences of the illness (appendix 10).

Analytic procedure

IPA involves four basic stages (Smith, Jarman and Osborn, 1999; Willig, 2001). First of all the transcripts were read several times and the researcher recorded initial thoughts and observations in the left margin (see example appendix 11). In the second stage the researcher identified and labelled the emergent themes that characterised different parts of the text in the right hand margin. These higher level ‘conceptual’ themes captured the essence of what was represented in the text. At the third stage the researcher attempted to create a structure out of these conceptual themes by looking for connections. Some themes clustered together while others emerged as super-ordinate headings. Each transcript was subjected to this process individually creating a preliminary list of themes (super-ordinate, main, and sub-) for each, with associated quotations to illustrate them (see example appendix 12). At the final stage the researcher looked across the lists of themes and produced a final structure containing the themes and the super-ordinate structure. At this stage some themes were merged, dropped or raised to a higher level.

N.B: In the interests of confidentiality, fictitious names were assigned to each participant and all individuals referred to in the interviews at the point of transcription.

FINDINGS

Twelve salient sub-themes emerged from the data and were clustered into four main themes, which were: ‘The opening of the eyes’ (Becoming aware), ‘The double-edged sword’ (Entering the system), ‘The adaptation’ (Becoming a carer), and ‘The maintenance’ (Surviving it). ‘The opening of the eyes’ (Becoming aware) and ‘The double-edged sword’ (Entering the system) were themselves subsumed under the super-ordinate theme of ‘Emergence and Realisation’; whilst ‘The adaptation’ (Becoming a carer) and ‘The maintenance’ (Surviving it) were subsumed under the super-ordinate theme of ‘Life Adjustment and Coping’ (Table 2).

Table 2: Table to show summary of themes

Super-ordinate themes	Main Themes	Sub-themes
Emergence & Realisation	‘The opening of the eyes’ (Becoming aware)	<ol style="list-style-type: none"> 1. Noticing changes: What raised suspicion... 2. Recounting landmark stories: What was serious... 3. Understanding: What might be going on...
	‘The double-edged sword’ (Entering the system)	<ol style="list-style-type: none"> 4. Getting labelled: What it is... 5. Getting help: What is out there... 6. Getting researched: What might help...
Life Adjustment & Coping	‘The adaptation’ (Becoming a carer)	<ol style="list-style-type: none"> 7. Reassessing relationship: What is lost... 8. Accepting: What is and what could be... 9. Readjusting: What needs to change...
	‘The maintenance’ (Surviving it)	<ol style="list-style-type: none"> 10. Managing daily life: What needs to be done... 11. Managing other’s emotions: What they feel... 12. Managing self: What about me...

Super-ordinate Theme: Emergence and Realisation

This super-ordinate theme refers to the participants' experience of becoming aware of the difficulties with their relative, their increasing knowledge of the symptoms associated with fvFTD, and also their interaction with statutory services.

'The opening of the eyes' (Becoming aware)

All participants were previously unaware of the existence of fvFTD until their relative was diagnosed. This theme describes the kind of journey that the relatives of people with fvFTD might be involved in to a greater or lesser degree. Participants talked about noticing problems, sometimes many years before finally receiving a diagnosis, and how they put these down to something else, and tried to understand them in terms of other explanations. Essentially though they felt there was something 'not quite right'.

This gradual build up of experiences and incidents commonly culminated in the participants recounting more specific and detailed accounts of major incidents, or what might be termed important landmark stories, at which time they clearly came to understand that something was 'wrong' with their relative. The journey inevitably continues, and as understood here involved a degree of information gathering in order to increase knowledge about fvFTD; either purposefully by the main carer or through other supportive relatives or professional carers. This led to a greater understanding of their relatives more individual and specific patterns of behaviour, even if it did still appear strange or nonsensical to them. This led to the difficult concept of coming to terms with the situation, which in this condition (and from these participants' perspectives) did not seem to be a clear-cut stage to be reached; the person with fvFTD is always changing, is expected to deteriorate, and in addition is also usually a treasured relative. The dynamic nature of the illness presentation and the difficulty making sense of the symptoms, together with the nature of the relationship and its deterioration, seemed to make this a particularly distressing and difficult process for the participants to undergo.

1. Noticing changes: What raised suspicion...

Participants described how they first became aware of the difficulties with their relative. Initially, these were often described as ‘minor issues’ and other explanations may have been brought into play to account for the patient’s aberrant behaviour, for example Mr Jones said:

R: ...when you noticed that things were going on you weren't sure...? Pt: Well no they weren't serious things then were they - I mean we were sort of thinking - oh you know "mum's going through the change". You know - it wasn't that bad you know what I mean... you noticed little subtle things...

(Mr Jones p.8, 429)

He also described how his wife’s motivation started to decline, which evidenced itself in her taking less care of her appearance:

...you know how women are - they like to look their best don't they...and she didn't seem bothered too much that way - you know what I mean - she wasn't bothered about her appearance....So things like that started suffering - you know what I mean...

(Mr Jones p.6, 266)

Mr Mills, on the other hand, became aware that his brother’s usual habits and interests were in decline, which caused him to feel suspicious:

Cars were his other passion. He worked on them. He used to take pride in them. He used to polish them rigidly weekly. And then... that was the first thing that probably triggered me thinking something was wrong, was he couldn't be bothered with his car, which was bizarre.

(Mr Mills p.42, 1967)

Mrs White described how the build up of a few incidents and mishaps, in the context of a very close relationship, alerted her that something out of the ordinary was occurring in terms of her husband's functioning:

He started having a few sort of memory problems and a bit of confusion... and we've been married for 37 years and we're very very close... and you know, you KNOW when the other person isn't right! ...erm forgetting like ... forgetting to turn the cooker off cos he'd come home put the tea on when I was at work. I'd come home and the oven was on fire, and well silly little things like that, like he couldn't find his keys but it was more than your norm, you know, and it wasn't right... and confusion, he was getting confused about things erm and overall I just knew.

(Mrs White p.1, 9)

So this theme encompasses the early experiences of all participants, the build up of curious incidents involving their relative which made them aware that something unusual was occurring; this ultimately led to them seeking further advice or help.

2. Recounting landmark stories: What was serious...

This theme encapsulates the seriousness of the illness, and the associated behaviours and symptoms. The theme includes what might be thought of as 'statements of extremity'. All participants recounted stories of when they had experienced something so serious or unusual that it clearly was not felt to be within the normal realms of human functioning and this was often the turning point in their own minds that their relative was seriously ill. They are stories that have been committed to memory. For example, Mr Jones described how he and his wife had both been smokers all their married life but she stopped one day and he felt quite perplexed that she never again mentioned it:

Well the one that really amazed me ...urm she used to smoke - well we both did - urm... I mean she was more of a smoker than I was... and urm... I'll never forget - I mean she was one of those people who would take a couple of cigarettes out of the packet and leave 'em on the windowsill so when she got up there was two there - and she could light one up straight away like - and then she'd have the other one while

she was having a shower or brushing her teeth or whatever, and she just packed it up like that! Never even mentioned it. She never even said “oh I’m dying for a cigarette”... Nothing. No sort of withdrawal symptoms – absolutely nothing. She just stopped and she hasn’t smoked for 4 years ...and not even said to me even once “ooh I’d love a cigarette” and I found that really strange that she could just stop like that... listening to how other people used to struggle with it and she seemed to do it so easily - so that was a major one for me – I could never quite understand how she done that!

(Mr Jones p.9, 460)

Mr Mill’s brother was forced to leave his own family and came back to live at his parent’s house at which point his curious behaviour became more apparent. Mr Mills described how his brother’s driving behaviour changed in conjunction with his ability to plan and manage his journeys:

But like I say it was the strange behaviour that came after it that really started and the truth is...really... initially... he rung me up one night, after we first realised that something was up, saying, “I need some money.” “Oh why, what’s the problem?” “Oh, I’m just standing in a petrol station, filled the car up with fuel and I’ve got no money.” And he knew he’d done it and he knew that he hadn’t got any money yet he proceeded to fill the car up. So, I sorted that out and then I spoke to my Mum and Dad two days later and he’d done the same again with them. “Well where’s all that fuel gone?” and he said “Oh, I’ve been to XXXX.” “Right, ok.” “But I’ve been there three times in two days.” It’s about... a hundred miles, something like that. So it’s quite a way. So he’d been three times in two days. He’d been twice in one day hadn’t he really, by the sound of it. (Pause) So that was strange...

(Mr Mills p.4, 187)

Whilst this lack of executive planning affected his own functioning it also began to impact on those around him. Mrs White recounted a story where this became even more salient as she feared for her husband’s safety as a result of his impaired work standards in the face of an irate customer:

This guy come over with a gang of his mates - wanted to punch my husband's lights out! Apparently it was a job that he'd done and he should have gone back to repair something, and I kept reminding him... I knew of the job and I kept reminding him and leaving little notes on the wall for him... but when he went out and started another job he forgets and this went on for a good long while – and this guy said “You're f-ing ripping me off!” and this, that, and the other, and he literally come right up to here with Brian. And I was pulling him off, pulling Brian off cos Brian wanted to bash him one! I tried to tell the guy that I'll sort it out – “Just tell me if you want compensation” I said “I'll sort it out... we don't need this... he's not well – we're going down the lines of him going into retirement” and he was like “That's a load of rubbish!” and I had to drag Brian off in the end...

(Mrs White p.5, 242)

Not only did she describe feeling threatened by this disgruntled customer but also she was aware that her husband's behaviour was inappropriate and risky perhaps reflecting a lack of self awareness.

3. Understanding: What might be going on...

The theme of 'understanding' refers to the experience of 'getting to grips' with the illness and its associated nonsensical behaviours and symptoms, either before or after diagnosis. It involves the search for explanations, for example in searching the internet:

There didn't seem to be anything then. You were just told, you know, the diagnosis... And then, get on with it basically... we didn't feel like we had any help... at all... Obviously we'd never even heard of Pick's disease... um... So then it was a case of us going on the Internet and reading the worst case scenarios. Well we had to! We'd never heard of it.

(Miss Green p.6, 268)

...as well as finding reassurance from, or receiving validation from a trusted medical professional or someone of knowledge:

R: Mmm - what about in the earlier stages... did she have some kind of insight...? Pt: She would always deny it... err... you know err... on the onset she was quite violent – and we'd never ever been a violent couple - but she was always attacking me - I could be sitting here watching the television and she would just get up and start to attack me! Trying to hit me – trying to pull me hair, claw me, what have you... and I remember it was one weekend, and I had to sort of put me arms round her to keep her arms down by her sides – to restrain her like you know...and she was still sort of struggling like - so I laid her on the floor... and within about 30 seconds she sort of went all relaxed... I sat back down and she got up and sat back down and held me hand – and I said to her “Why did you do that?” and she said “Do what?” and I says “Well - you just attacked me!”, “I haven't” she says “I haven't”, I says “You have”. She had no recollection of it! I remember Dr Foster saying “Yes... they can be a bit like this - especially with the onset” ...and that sort of violent streak lasted for about 6 months then it stopped, and it's never happened since.

(Mr Smith p.35, 1810)

This theme also encompasses the participants' own attempts to generate answers directly from their relative. Mr Mills described how he tried to understand his brother's behaviour by asking him direct questions. He discovered that by doing this he became aware of his brother's circular thinking, describing it as a vicious circle:

When we used to ask about, for instance, the food problems... “No, I'm not touching that, not touching that”. We used to ask him, “Why do you need to eat, John?”, “Well I don't”, “So, why are you eating?”, “I don't know” ...and then erm... he'd eat a meal, and we'd say well, “John, why do you feel the need to walk it off?” “All the adverts say so”, “What adverts?” No answer. “Well they tell you to walk ten thousand steps a day, right. So I walk round the block and come back and eat.” “Right, but try not eating John.”, “I know but I'm hungry”. And it become like a vicious circle. And you couldn't break that cycle. No matter what was explained to him.

(Mr Mills p.9, 416)

So whilst this theme contains the participant's strategies, techniques, or attempts to try to understand or make sense of their relative's behaviour, also subsumed within it, is the participants' ability to actually accept and work with the 'strange' behaviours; showing an understanding of what is needed in order to get by. Mr Smith describes how his wife had lost her sense of humour and how one needs to be aware of this, and careful what you say around her, in case she takes offence. Whilst he is able to manage this in private, when out in public he is not as able to manage other people's interactions with her, for example:

...she doesn't have much of a sense of humour now although she did have up until about a few months ago – you have to be very careful what you say when we're out and about - if we meet people we know they have to be careful what they say - because she takes offence very easily now... somebody the other day for instance - you know - we seen them when we was over the local shop and... err... the woman says to me "Hello Robert" and then she said to Maggie "...and are you alright Bab?"...and well that is offensive to Maggie now you know - and she says "Did you hear what she just called me? She's just called me Bab!!" And now she makes a big thing and big issue out of it... it's very very strange.

(Mr Smith p.20, 1023)

...and Mr Jones also evidenced an understanding of his wife's condition, and the changes occurring along the way, and responded accordingly:

Six months ago she would've probably... well maybe a bit longer than that... she would have been asking me to take her to town... but you can't - it's really a waste of time because she never used to shop. I just thought that she just wanted me to take her to the shops so she could just ...steal... and if I showed her something, like "Do you like this dress?", she just wasn't interested. She'd say "No" and just walk off! So it soon became just a waste of time doing stuff like that - even though I still take her food shopping.

(Mr Jones p.22, 1145)

Finally, Mr Smith also talked about other people's interactions in the sense of how they perceive his wife. Other participants were also aware that the illness is relatively invisible, which can lead to misunderstandings or difficult interactions:

...and when I'm out and about with her now – because she's so jolly and she looks physically well - people always say "Well she looks fine to me, there doesn't look anything wrong with her" and I says "Well it doesn't show on the outside you know" ... So err... when they say to her "How are you Maggie?" and then all of a sudden she grabs hold of them, you know, by the shoulders, in a friendly sort of way... but they're a bit sort of... 'well what's happening here...' sort of thing! And she'll say to them (and she'll say the same thing over and over again you know) "What I say" she says "Carry on regardless! Look on the bright side!" And she tells everybody that you know [haha] ... so they all say "That's right Maggie - that's right!" ... but it's when she grabs hold of them you know – she would never do this before - you know - but now it could be a complete stranger...

(Mr Smith p.36, 1874)

In this story Maggie's behaviour also evidenced some lack of understanding about other people's feelings, or diminished awareness of personal space, which may be a result of a deterioration in her understanding of theory of mind – that different people can hold different views of the world - and it also indicates a lack of empathy or understanding on her part, perhaps relating to the difficulty of reading the 'mind in the eyes'.

'The double-edged sword' (Entering the system)

All participants' relatives, through developing fvFTD, had become involved with health and social care services to varying degrees. For most participants this involved an initial assessment with the GP followed by a referral to more specialist services, although as will be discovered this was not necessarily a linear and clear cut process. Seeking help for their difficulties by entering the statutory service system was not a straight forward process. Whilst participants recognised the need for help and reassurance, they found they had to negotiate a complex system where adequate and appropriate service provision was lacking, and knowledge of the specific nature of the difficulties and the needs associated with people

with fvFTD was patchy. This experience could then be described as a ‘double-edged sword’, since, on the one hand, the participants were in need of information, support, and treatment but by entering the system in which they hoped to receive these, they often found limited provision, if anything, which also involved long waits, uncertainty, and lengthy bureaucracy. One participant described this as ‘like negotiating a minefield’. Eventually it seems that once the initial assessment (both medical and social) had been undertaken and a suitable service had been located and provided then participants felt more connected and supported, and stress reduced.

Throughout the following three sub-themes, the ‘double-edged sword’ is evident to different degrees in a dimensional sense, which changes depending on the participant’s personality or temperament, the stage of the illness, and degree of difficulty experienced, in addition to the nature of the problem encountered. The kinds of feelings that came across in the interviews were: hope versus despair, isolation v connection, loss of power v power, vagueness and uncertainty v reassurance. There was a sense that in order to receive the benefits of the system, one needs to be ‘at the mercy’ of the medical system, giving up power and relying on others – and sometimes this seemed difficult for participants to accept.

4. Getting labelled: What it is...

In order to try to understand what was happening to their relative, participants described seeking a diagnosis. For some it was a long and difficult journey with many meetings and assessments, much waiting, and the experience of uncertainty. For example Mrs White spoke about this:

We’d seen a couple of psychology-type things – one of them in the Hospital and he said that, well he didn’t mention dementia but he said it was some sort of memory loss thing and he said it could go - you know - stay as it is or a middle pattern or it could go right down quickly or it could go down slowly - yes and he thought it would go down slowly – this was maybe 4 or 5 years ago.

(Mrs White p.3, 111)

...and Mr Smith also described the process they went through, which involved a misdiagnosis and a period believing that his wife had Alzheimer's disease, before a more appropriate diagnosis of fvFTD was ascribed:

....we saw Dr Ahmed several times down at the Psychiatric Hospital... at first he sort of said it was Alzheimer's disease... and... I think for about 18 months he thought it was Alzheimer's disease - and then the one visit we had with him he said "I think that I might have got this wrong"...and he organised a special sort of scan... I'm not sure what it was called but they injected Maggie with some sort of radioactive material...and then did this scan and then eventually he came to the house and he said "Right, we have got the diagnosis now!" which was frontal temporal lobe dementia. So that's how we found out what she's got - but it took about 18 months to just over two years before we got there in the end.

(Mr Smith p.2, 81)

Two participants, Mrs White and Mr Brown, were very dissatisfied with the service they received. Mr Brown's GP appeared to seriously lack knowledge, not only of the pattern of symptoms but also of the mental health and social care system. He felt compelled to seek help outside the National Health Service and turned to the private sector. He also talked about how important it was for them to find out what was going on in order that they could adapt and prepare. Unfortunately, Mr Brown's dialogue here also very clearly indicates the 'double-edged' nature of receiving a diagnosis:

Obviously you want to know what's happening don't ya, as soon as possible. So I mean you wanna know so you can adapt... so, yeah, in that respect... The frustrations came after the diagnosis, I mean, well obviously, when...we got the diagnosis, I was very disappointed, cos I mean you... phfff. You hit an all time low when you hear the news. [to the patient] And you were a bit...er...you walked out the room didn't ya when he er, when Dr Small came... Do you remember? Very upset. Very upset - well I was upset as well, I don't mind telling you that, obviously.

(Mr Brown p.26, 1331)

Mr Jones discussed how receiving a diagnosis of Pick's disease impacted on him at the time but also how having an explanation for the cause of the problem still affects his thinking now, again demonstrating the bittersweet nature of knowing:

Well it was the consultant at the hospital – Mr Markum – and I mean he said to me like “Your wife’s got Pick’s disease...”, “What’s Pick’s disease then?” And urm... obviously its frontal lobar dementia and erm... there’s no treatment and there’s no cure! So like at that point I was.... I was in shock! I think you could have just done that [does action] and I’d have... fallen over! I just didn’t know how to take it – and I think from that point I’ve sort of been in a - well I don’t know how to explain urm I mean its what’s been told to me you know ...but it’s like a sort of... well I can’t take it in...strange innit?

(Mr Jones p.11, 540)

5. Getting help: What is out there...

The barriers encountered in trying to access services in order to gain a diagnosis and find out what is going on have already been touched on but this did not stop with first contact with services. Due to the rarity of the disease and it only just getting recognition as a distinct disorder, requiring specialist assessment and treatment, health services and health care workers of all levels are ill-equipped to deal with it. First of all Miss Green clearly states from her experience that, although getting a diagnosis was important, there did not appear to be enough follow up support, she said:

I think when you initially get the diagnosis I think you should have more support then, sort of. Somebody to tell you exactly... what it is and what’s out there, what help’s available. Cos it didn’t seem like we had much help to start with. It only seems like now that we’re getting help and support.

(Miss Green p.28, 1313)

It might be the case that her family now know of the avenues of support available with the benefit of time, but other participants’ experiences, indicate that initially the knowledge or the infrastructure is not offered. It is almost as if each participant presenting with their relative to

services is a 'test case' that requires the service to then develop services around that person. At the time of interviewing Mr Brown still had not received any support even several months after the initial diagnosis:

So we went back to Dr Stansted... and he said, "What local help are you getting?" And we said "We got nothing!" Six or seven months – nothing! Even to this day we haven't actually got anything – nobody – have we?

(Mr Brown p.4, 206)

He also went on to explain why this had occurred, again highlighting very poignantly the 'double-edged' nature of entering services:

So of course we come back all enthusiastic [from a visit to a good suitable care home] "Yes, yes, yes, we're pleased with this" ...and then he says "Ah well, I can't get a social worker appointed to her until January..." He says "I could get a local social worker from the area – but they don't do the same things as us – well they're not under our umbrella... they don't do the same things as we do – they mainly deal with over 65s whereas we do under 65s, we lose some control of the patients really... so I would rather ask you to wait until January, if you would, until we get the proper person appointed and then they will come under Professor Packard's umbrella and there's a group of us..." (I think there is about 7 or 8 of them in the group; nurses, social workers, and care workers). He said "They do... well they know the places to go, they know who they use... and it will be much better" he says "If you can wait..."

(Mr Brown p. 10, 520)

Another barrier experienced by several participants, which caused frustration, was the age appropriateness of services available. From the participants' experiences it seems apparent that respite and residential or day care facilities are set up for older people with dementia, people over 65. Participants experienced either a dilemma related to envisaging their relative in a care home for the elderly, as Mr Mills puts it:

...we certainly didn't want him in an elderly home, you know ... you can imagine it can't you. You can't stick a forty year old in a room full of people like that - through no fault of their own, dementia patients, but for a forty year old to live in an eighty year olds' care home.

(Mr Mills p.37, 1765)

...or they were rejected by the service because they were under the age of 65. But not only were they rejected, as Mr Smith puts it, they were led to believe that it was available only to be turned down further along the line, again reflecting the 'sting in the tail' associated with services:

They've tried to fit Maggie up with another sort of day care centre ...and basically the one - she spent the day there you know - and they said she was ok - you know - and that she would be acceptable... and then they said "We can't sorry, she's under the age of 65!"and this is what you find all the time. So if this closes at the moment I've got nowhere else for her to go... She's 61. And well a lot of them would be older... I don't know, yeah. It's pretty standard I think everywhere they've tried for her - you know - under the age of 65 so... Yeah it is annoying – it seems to be that some people probably think that people under the age of 65... don't need that ...no... they don't get dementia.

(Mr Smith p.6, 267)

6. Getting researched: What might help...

All participants had some experience of research, either because they were taking part in the current study, or because they had at some point been made aware that they may be able to enter a clinical trial of medication. In fact none of the participants' relatives were at the time of this study enrolled in any medical research, however one participant's relative had just been prescribed the trial medication, Methylthioninium chloride (Methylene blue) 'out of trial' with funding from the local Trust. The possibility of a medication that may help was found to be encouraging to those participants offered it; giving them hope that something may come of it. However, the promised trial did not materialise leaving the once hopeful

participants languishing at home feeling helpless and let down. Mr Jones summarises his experience as follows:

He said to us that he was running some trials – he wanted to trial this drug... So we went and met Dr Fox... and he says “Yeah, I want...”, he ...well he looked at her and seen how far she was... and how she presented herself and he said “Yeah, she’ll be part of the trials when they come about!” and well that was two and half years ago! And those trials were gonna happen that year, but since then we just keep contacting him and saying “Is there anything happening with these trials yet?” He gave us a lot of hope urm... whether that was a good thing or a bad thing I don’t know... we booked another appointment to see him in another 6 months... and he said well hopefully they’ll be something happening by then... But we’ve heard this for the last two and half years but - you know - we have to keep going...

(Mr Jones p. 34, 1762)

On the contrary, Miss Green’s relative, who was eventually offered the trial medication, tries to describe her thoughts and feelings around this. Sometimes she seems conflicted and uncertain about the benefits or implications, but overall is grateful for the opportunity to try something:

She’s started some drugs now. That’s been another thing, I think in the back of your mind you sort of... think that she’s gonna get better... and maybe that’s what keeps us going... the fact that she’s taking these tablets you think that one day she might get better, or she’s staying at the same stage because of the tablets. And that’s something we’ll never know cos she’s not part of a trial, it is just... my Mum that’s taking them. We’ve been told that it won’t make her better and that there’s only a, I think it was a 20% chance that it could keep her where she is or slow things down. We couldn’t have NOT given her the tablets knowing that there’s something out there, you know. We’ve got to try it haven’t we. And I think that’s another thing psychologically, I think sometimes you think “Oh she’s done that and she wouldn’t have done that before.” Do you know what I mean? I think it makes you think

things... you know... sometimes I think you kid yourself that err, she's getting better because you know she's taking the tablets.

(Miss Green p.24, 1110)

Super-ordinate Theme: Life Adjustment and Coping

This theme refers to the participants' requirement to respond and make changes both outwardly and inwardly to the demands of their situation.

'The adaptation' (Becoming a Carer)

In all cases the participants expressed, through their stories and their language, their development or shift into becoming a carer for their relative. This theme includes issues related to the shift in relationship that necessarily occurs as the person with fvFTD functionally declines and exhibits more behaviours that require understanding, management, and acceptance. But not only this, fundamentally as the person with fvFTD becomes more affected by their illness then the symptoms begin to alter their personality and the way that they interact. Participants talked either openly about their experience of becoming more distanced in their relationship, or it was evident in more covert ways essentially changing the dynamics between them. This theme therefore also draws on the experiences of participants in how they come to accept their new role, by recognising the pressures inherent in it, not only through the loss of their usual way of relating, but also in needing to live with the uncertainties. There was also a sense that the situation becomes 'normal'. Finally another subtheme here is that of the need to make adjustments and adapt to the new role of carer, which may mean giving up things (such as leisure pursuits) or developing new skills or qualities (such as a sense of humour).

7. Reassessing relationship: What is lost...

This important sub-theme, as already suggested above, draws on participants' accounts of their relationship with the person with fvFTD and how the illness changes the way that the person with fvFTD interacts. Essentially their personality alters, and this in turn alters the way in which they can be related to. Mr Jones talks about this, he said:

Well I mean it is like what I was saying to you - the person that I'm caring for now is nothing like the person that I married - you know what I mean – she is different and that's what hurts really... I mean - it's the same person but all the qualities that I saw in her have gone now you know... all the personality has gone.

(Mr Jones p.32, 1659)

Mr Mills recounted how his mother had described her feelings about her son with fvFTD:

It was a big culture shock to them. Their eldest son (pause)... as me Mum described it first... "He has been taken away, by something, and I can't get him back!"

(Mr Mills p.27, 1278)

Miss Green described this as like 'losing her mother'. One of the important things that she missed was the fact that she could no longer have a conversation with her, she said:

She doesn't talk a lot at all these days. She sort of copies what you say so it just means not having her there to talk to, and it feels like she's not here, because she's only here in person but not in mind...

(Miss Green p.3, 132)

Mr Smith described how, at the time of the interview, he had experienced some recent situations in which he had been made aware that his wife was losing her connection with him. He said that she had asked him one day in the kitchen if he knew when her husband was coming back. When he replied that he was her husband there was no response. He also said that on several occasions she had checked with him what his name was:

...and there's been other times we've been sat here and she'll say to me... err... "Your name it's Robert?" And I'll say "Yes, it's Robert" and she'll say "Well I have to keep reminding myself what your name is..." so yeah you know it is... it's strange... and sometimes it's heart breaking you know...

(Mr Smith p.19, 993)

This not unsurprisingly caused him some sadness and distress. Mrs White explained her experience in losing her connection with her husband quite graphically and emotively, indicating how she was struggling with the change in roles from being a partner to being a carer, she said:

I'm a carer, a mother, a nurse. I'm treating him like a child three quarters of the time and then I've got to reverse my roles to be wife again ...and a lover and one thing and another... erm I have to keep my eyes closed cos if I open my eyes... and see his face... I'm seeing this face that isn't him... and it just doesn't feel right... to be doing the sex bit. I love the cuddles... We always cuddle each other and when I wake up in the night I'll put my arms around him and its great! But to go... well the other bit... I wish he would go off it to be fair... I mean it wouldn't mean that I didn't love him any the less because of it...

(Mrs White p.38, 1980)

8. Accepting: What is and what could be...

In transition from the role of partner (or close family member) to being a carer, individuals began to recognise and accept their new status. Mr Jones said that one of the situations he was in really brought this home to him, in a consultation with his GP:

The GP she said... "Have you had your flu jab?" And I said "Well, no, I'm not eligible am I? You have to be over 65, don't you? And she said "Oh no, you're a carer... you can have your jab now" which I did. So you see stuff like that...

(Mr Jones p.27, 1420)

Whereas Mr Smith was in receipt of regular respite care and had managed to develop certain ways of coping despite feeling the pressures brought on by his wife's behaviour. He said:

R: So it sounds like over the course of the illness there's things that you've learned to cope with... certain ways of... with the things that have changed? Pt: Yeah - I'm sort of very sort of ... annoyed at times with her, although I never lose my temper with her - you know - erm... because she's very trying, you know, and when you've had it

all day long and it could be ten o'clock, half past ten at night... she'll go to bed - I'll stay up another hour and then go to bed then and as soon as I get into bed she'll start with this behaviour again you know...

(Mr Smith p.26, 1349)

Finally, another aspect that came across quite strongly in the interviews, and is an integral part of the acceptance of the new role / situation, is the sense that through finding themselves in unusual situations, and experiencing and dealing with the 'strange' symptoms associated with the illness, somehow this becomes normal to them, as they become accustomed to it:

She does all these strange things - you know - I've become sort of used to it - accustomed to it - and I don't draw attention to it - you know - she'll put two night dresses on to go to bed and a work's overall from when she used to go to work... but you don't make a big thing out of these issues, you know... I do tell her... but I don't sort of argue about it - there's not much point... I've learned to sort of cope with these things... well she says "Well it won't hurt - it's up to me what I wear" so I say "ok fine if that's what you want to go to bed in that's fine"

(Mr Smith, p.26, 1328)

When out in public, Mr Jones described the following experience where he has learned to accept other people staring at him and his wife, he has accepted this reaction from the general public as normal. Not only has he realistically appraised the situation, he has also normalised it and although it bothered him at first, it no longer has the power to do so:

But like everybody looks at her now cos I guess people perhaps recognise her now - and perhaps think "Oh, she's here again this mad woman..." Well - you know - I think you would wouldn't you really - I think if I had that experience and I saw someone coming and shouting, your reaction is to turn round and look so the reaction that she gets I would think is quite normal - I would think - but then it just draws attention to us all the time... that used to bother me a little bit... yeah - it don't know, no, I just think "Right, well that's how it is!" you know.

(Mr Jones p.23, 1192)

9. Readjusting: What needs to change...

As part and parcel of taking on the role of a carer for someone with fvFTD, certain changes seem to have been necessary for the participants, depending of course on their individual situation and circumstances. Mr Brown talked about ‘adapting’ several times in his interview, and part of his adjustment has involved him taking on the lead role and responsibility for organising and carrying out domestic chores:

I cook everything, I think I’m a dab hand now... (Laughs) I cook everything and weekends I try to do as much housework as possible but it is difficult I mean, I tend, if I’ve got the downstairs here pretty clean, the upstairs suffers, you know. We have to keep it... it’s difficult to do everything... Err ...all the washing, all the ironing... I do all that... so... err... yeah, I’ve adapted!

(Mr Brown p.21, 1087)

Other participants talked about how they had had to give up leisure pursuits and hobbies, facing the reality that they do not have enough time to commit to them while caring for their relative. Mr Smith talked about his experience:

Most of my time when Maggie is here is taken up with Maggie all day long you know... well I used to have fish in the garden I haven’t got that no more now - the garden’s over run now - I don’t get so much time to do what I used to do in the garden – even in the house - you know - it’s difficult... I used to have big Koi carp out there - big ones... and well I gave everything away about two year ago now – all the fish, all the equipment - and I’d kept fish for about 25 years – I just couldn’t get the time to sort of do what I needed to do like to look after them – I just couldn’t do it...

(Mr Smith p.22, 1113)

The issue of employment also featured heavily in participants’ accounts, some taking early retirement to take on caring responsibilities, but others not able to do this and finding themselves juggling work and home care commitments. This is an important area since people

with fvFTD are generally of working age and correspondingly their partners and carers also. However, Mr Jones was able to fortunately take early retirement:

I was in engineering. So I was like a supervisor in engineering – I'd been with the company for 38 years – so I err obviously I err well somebody had to look after her so I retired early. R: ...so things changed quite dramatically...? Pt: Yeah, yeah... So from doing urm... well sometimes weekend working like shifts and working all my life – all of a sudden just stopping...

(Mr Jones p.13, 677)

Another aspect of life as a carer is the possibility that one may need to develop, not only skills, but personal qualities that perhaps had not been so important before. For example, Mrs White described how she had been able to draw out a 'sense of humour' in order to best manage the difficult situations in which she found herself with her husband:

Brian's always been the laughy jokey barmey type... he's always the clown... he can get everyone laughing you know... and I was always the shy one sitting in the background... well I've confidence in my shop while I'm in it yeah but anything out there... put me in the background you know! And I hadn't got a very good sense of humour at all... But I don't know how or why I did it butwith the things that he was doing or saying and still does now... what choices I said to myself have I got? I'm not a shouty yawpy telly offy type person – I don't like conflict and things like that I can never belittle him and put him down – and I can't keep telling him "No, you've done that wrong", "No, don't do that" So somehow or another out the blue it come, that if he was doing something that was wrong or whatever... I would turn it round as a laughing joke... and then we'd both end up laughing about it! And I don't know how I started doing it or what gave me the insight to be able do it... but that way works best for both of us.

(Mrs White p.40, 2101)

‘The maintenance’ (Surviving it)

This main theme details participants’ methods of coping and surviving with their relative’s illness; the way they live with it day to day. It includes what they have had to process internally, in terms of reconsidering their own lives on a practical and emotional level, and the skills and techniques they have had to draw on, and put into practice. Adopting the new role of carer involves ‘managing’ the person with fvFTD on a daily basis and keeping them occupied. It also involves taking into consideration their emotions, thoughts and happiness – considering their humanity. A big responsibility is the need to take on the management of other family members’ emotions, in particular in terms of protecting them from the reality or the extremities of the disease and its symptoms. Finally, through their use of language and expressions and the way that they communicated, information on how participants may manage their own emotional wellbeing throughout this process was conveyed, and related to this, some more practical ways in which this can be achieved are evidenced by some participants.

10. Managing daily life: What needs to be done...

Participants talked about different symptoms and behaviours that they experienced regularly, and the techniques and methods that they had learned to employ to manage these situations. It seemed as if participants began to develop a repertoire of these strategies in response to the demands of the role. A major part of caring for someone with fvFTD involved supervision, and Mr Jones described his need to prompt his wife to carry out activities due to a decline in her self-motivation, he said:

R: You sort of prompt her to do things then? Pt: Yeah. I mean like “Brush your teeth”, “Put the toothpaste on the brush”, I hand her the toothbrush and she’ll brush her teeth - and I give her a brush to comb her hair, and she’ll have a little go at it - but everything is half-hearted - you know. She won’t do them properly. She just loses interest - there’s no drive there.

(Mr Jones p.24, 1253)

Another reason for supervising the person with fvFTD on a daily basis was related to safety. Miss Green talked about how, in the earlier stages of the disease, her mother had left the

house on a few occasions and driven off in the car causing some concern. In response to this the family had increased their supervision of her:

My Dad can still go and do gardening whilst she wanders around the house as long as the doors are locked and the keys are hidden. Then he can still do things like that but obviously you've got to be with her all the time. You couldn't, sort of, you know we couldn't leave her in here and go shopping, you know cos you've got to... cos she has got to have somebody with her all the time.

(Miss Green p.18, 813)

Mr Mills described another aspect of safety in relation to his experience with his brother. He found as part of his caring role that he had to consider the risks that his brother's behaviour triggered:

We were trying to find out what we could... you know, how we could treat him, or where we could put him in a place of safety or anything. Cos he used to go to the local park... sit on the swings at school time when the kids were coming out. Speak to them and we didn't want people misconstruing... having ideas, you know He was poorly. We recognised that he was poorly, but other people didn't.

(Mr Mills p.8, 371)

Mrs White talked about how she had needed to develop an 'anticipatory strategy' while carrying out daily tasks and activities in order to avoid mishaps:

...and the clumsiness he's had that right from the start but it's getting a darn site worse now – it's like having a child – mmm – if we're in the kitchen bit there and the same down stairs – I know it's not big but if he's going to get a glass of water and I'm doing a slice of toast, and we're both coming the same way then I have to side step, I have to watch, you know, I have to keep out of his way you know cos he'll have me over. If he's got a cup of tea I have to be very careful because he'd spill the cup of tea, and it could go on me, and all the saucepans and anything with a handle

has to go right in on the cooker just like when you've got children, you have to anticipate what's going to happen, yeah, you do have to have fore thought exactly.

(Mrs White p.34, 1785)

Both Mr Mills and Mrs White talked about how they had to employ a certain amount of manipulation in order to manage their relative and essentially keep them safe. Both found telling 'white lies' a useful and important strategy at particular times:

He used to have in his mind that, he wanted to go home. He wanted to go and see the kids or he was going to see Barbara, which was his fiancée. And that was the pretence when he first came down here. And we said, "John, you can't. You've got to stay here to get better." Arhm. We always said that, you know, "You're going to get better."

(Mr Mills p.23, 1057)

"I'm having a bike! I don't care what you say, I'm getting a bike!"... and oh my god a bloody bike! I mean he's bad enough in a car, a bike's gonna be even worse and I thought oh well he's put his foot down now and he's getting stroppy, let him get it, I can always find ways of wheedling round him, you know "Don't go out on it you know it's raining" or "John's gonna pick you up" or "Your son's gonna pick you up or..." so all right yeah he went and got this bike and he got the crash hat... I was just dreading the first time he went out on it – I watched him going down the road on it like this, and I was just like "oh my god!"... So erm the next time he went out on it – I kept making excuses from then on in, any excuse I could think of, whenever he said he was gonna use the bike I would come up with a better answer for why he shouldn't. I can't remember all the little lies I told him but fingers crossed and touch wood he hasn't been on it for a month or two now.

(Mrs White p.13, 654)

11. Managing other's emotions: What they feel...

Another salient sub-theme emerged from participants' experiences, and seemed to be an important and influencing aspect in the role of 'carer'. It revolved around the consideration of other peoples' feelings. Mr Brown's wife was in the earlier stages of the disease and he was still employed and therefore out of the house all day. He felt there was a need to check on his wife during the day but had mixed feelings about the impact of this on his wife's emotional health:

The biggest part of it now is that when I'm at work during the day... she's at home. She can't drive anymore - she had to give up her driving licence obviously. So she can't go out without anybody coming to take her out. I'm at work all day, and I can't afford to retire yet. I've looked into it but there's nothing, well not enough in the pension pot... to be able to do that, so she gets really down – really really down, during the day. I mean, I ring up two or three times during the day don't I? From work... to see how she is – and sometimes when I do that it actually makes it worse, don't it? Because when you hear my voice you want me back home, and then I can hear her voice change. But I mean, I still want to ring to find out how she is, but when I ring her... she's gonna be clock watching then until I come home. "What time you coming home?" Oh yeah, I might ring a number of times and I might not be back until half past four, and you know it's a long stretch to be waiting...

(Mr Brown p.6, 284)

Mrs White also felt she needed to consider her husband's feelings as part of her management role, in terms of damage limitation:

Yeah you do have to have forethought, yeah exactly. Because the thing is that even if it doesn't harm him the thing that he is doing – if he's knocking things over or breaking things it gets him more upset, so if I can... prevent that accident then you know it's better all round... I just need two heads and two brains to do it with [haha].

(Mrs White p.35, 1816)

Thinking about other family members' feelings in response to the illness in their relative also played an important part in the primary caregiver's life. Mrs White described trying to manage her children's experience of their father:

To start with when he first got bad I was telling them... most of the progress, like we've seen a doctor and they've said he should have that test, and then I'll say "Ooh your dad wasn't very good today erm he was erm forgetting more words..." or "He was breaking things up..." thinking that it's their dad and they should be knowing... you know, but I've stopped doing that now... my daughter especially, you know, "...every time you say anything to me it's doom and gloom!". I love em to bits, they're good kids and they've caused us no trouble at all - and I know they love us but especially me daughter she's a business woman... very very busy... she hasn't really got time... we probably see her once every two or three weeks and I'll phone her once a week and that's about your lot - it's obvious she doesn't want to know... she says "You'll tell me anything I need to know" and that's it, but then...again I blame myself because I've purposely tried to not put too much on them and shield them... yes, protect them a bit from it like.

(Mrs White p.24, 1253)

...and Mr Mills also felt responsible for his parent's emotional experiences:

I come down and have meetings with the staff and erm... I convey the facts that I get, in a watered down form to my Mum and Dad. I try to protect them as much as I can. I protect them really as much as I can because they don't need to know the ins and outs of what John's doing.

(Mr Mills p.31, 1478)

12. Managing self: What about me...

This sub-theme includes emotional awareness and practical ways of managing the self in response to becoming a carer. On the whole participants seemed able to recognise their own emotional state, their difficult feelings related to their situation. When asked about the most difficult aspect of his life at the time of the interview Mr Jones said:

...looking back. If I do - then that brings me down... too much. But if I just take each day ...and just work with what you've got then it seems alright... but occasionally - like if you're on your own – sometimes I start to look back and that's difficult because then you can actually see how far you've come... and that's... well I don't like doing that really too much... you know... looking at photographs if you like or... well I wouldn't say that I don't like doing it - but it depends – I know it's not going to do my morale that much good really.

(Mr Jones p.25, 1285)

...and Miss Green admitted that on the road to accepting the situation she recognises that there are times when things can become a little overwhelming:

And then obviously, there, there are times when you do break down and ...you know... have a cry and... let it out, um, I mean obviously you can't stay strong all the time, can ya? But, I'd say, yeah, over the past 6 months it's certainly been a little bit easier to... accept.

(Miss Green p.24, 1094)

Other participants also echoed these remarks, that there are times when it is not possible to keep up the veneer of strength. Mr Smith talked about his life as if for him it was like being on a rollercoaster:

*She doesn't like me laughing - but sometimes I'm in stitches you know! [Haha]
There's been some funny things you know! So - you know - it's laughing and tears at times. R: ... a bit of a rollercoaster...? Pt: Yeah, oh I've cried you know – some days when I see her at her worse - you know...*

(Mr Smith p.41, 2113)

In order to manage their emotions, participants seemed to have developed some strategies. Not dwelling on the problems and challenges that they encounter, and getting on with daily life, seemed to be a prominent method. Miss Green said:

And like me and my Mum were really close. We used to do a lot of things together... um... and it sort of meant we stopped all that because we couldn't really do those things anymore. You know. Now it's me who's taking my Mum out, you know, we're not going out together, you know I'm like... I'm like, looking after her you know it's difficult, but... you just have to get on with things.

(Miss Green p.4, 177)

...and Mrs White described very clearly a technique that she has had to adopt in response to a specific symptom exhibited by her husband:

Yeah definitely the gobble de gook is erm, I wrote a load down cos I couldn't, well like I said to the psychologist and Dr Churinga, this is how I dealt with it to start with, and I think I'm still probably dealing with it a bit like that now... it's if he said something that... well it's all upside down inside out it don't make no bloody sense - part of what I'm saying to you is that I try to put these to the back of my mind. When it happens deal with it then kick it out! Don't keep it in your mind in memory because he's gonna do another and another one and it's just - my head would get full of em! So I have to try and kick em out - so then if someone asks me to explain them I find it hard.

(Mrs White p.29, 1502)

Developing a sense of humour also seemed prominent for some participants. Mrs White's development of a sense of humour has already been documented, there follows an example:

We were going into a supermarket shopping the other week and just as we were going into the door he'll think of summat, erm, and he'll say erm ... "Underspray!" right "I need underspray - I want underspray - I need underspray!" and then cos... well.... it's hurting in here, but we joke - I've learned to get a sense of humour which I never had before and I say "What you on about you daft sod - underspray??" "Is it underspray for the car? Is it underspray for the bed?" or well you know I was being barmey you know [haha] no, no, and he got it then you know, under ARM spray...

(Mrs White p.31, 1591)

Mr Smith also used this humorous quality to help him cope, and it was evident throughout the interview, for example:

She became quite forgetful and she had developed a lot of strange habits and a lot of strange behaviour, but as time has gone on she has gradually got worse. So this is where we are at the moment...we're [haha!!] I always say that she drives me up the wall and by the night I'm walking on the ceiling [haha!!].

(Mr Smith p.3, 108)

Another element that came through during the interviews was participants' ways of managing their own shifts in emotions as they spoke about particularly troubling or difficult events. Here is an example from Mr Smith's interview, where, when talking about something difficult, he trails off and wonders aloud whether what he is saying is important or interesting:

They used to visit a bit more often and it's only since their mother has had this illness that they seem to keep away a bit more you know... she's just very very trying... she gets very confused with the things she does and says – she gets all the children's names mixed up! You know... erm... it is... it's awkward ...erm... (pause) ...well I don't know if you want me to ramble on the way I am...

(Mr Smith p.18, 920)

Finally, another way of managing their situation and emotional responses seemed to be to see themselves as being fortunate by comparing themselves to others. For example Mr Jones talked about another family that they knew of who had a genetic form of frontotemporal dementia:

But I mean their plight was far worse than ours really... you know - cos she had lost her mum when she was something like about 4 years old... and because her family was young... her sister had to look after them... and she later died of the Pick's disease... and like then her brother's just gone as well... and I thought well that's devastating that isn't it... So straight away - I mean - my situation is nothing compared to hers.

(Mr Jones p.33, 1738)

On a more practical level, participants talked about a range of activities which they could draw on to help them to cope with their situation, or things that enabled them to recharge their batteries or keep them motivated. Mr Smith talked about music being a great stress reliever:

R: ...so how do you cope? Pt: I've always said that I love me music - you know - I always have done – all sorts of music you know... many many times I still do – I did last night – err... when I'm feeling sort of really stressed out... I'll say to her "I'm going to leave you watching television" and I sit where you are now and I put me headphones on put a CD on and listen to it, and that way I feel myself coming down... Yeah – I find music a great stress reliever.

(Mr Smith p.27, 1396)

...and Mrs White was thankful for being able to retire to a nearby holiday caravan each weekend to unwind with her husband:

So – it's just the weekends can be a little bit of a problem but thank god for 10 months of the year we've got a caravan now... we bought a cheap one... and he loves the fishing and the water and peace you see... and because you can't have weeks off with the business, we go every weekend, we go Saturday afternoon and we come back Monday.

(Mrs White p.28, 1451)

Time away from the person with fvFTD also featured prominently in participants' accounts, either because they already had formal respite arrangements in place or because they were struggling to get this actioned. Mr Smith was fortunate to have been able to benefit from respite care for his wife but he described it at first as being a difficult adjustment to make, having spent many years with his wife:

I mean when she first started going in the first couple of times I found it quite stressful myself you know - cos we've been married for such a long time - you know - and all of a sudden that person is... as I say - the way she is terrible at times you still miss her... but the last time she went in it didn't bother me at all and I felt really

relaxed last time ...saw my brothers... and they phones and said “Well - we’ll come and pick you up - spend the day over with us” like, so they came, took me over to their area and we’d go and have a drink and a pub lunch, something like that... which is something I didn’t really get to do... and have a walk around somewhere - like you know - and I haven’t got to worry about getting back... Yeah - it was really relaxing the last time she was in hmm yeah.

(Mr Smith p.44, 2293)

Miss Green and Mr Jones both felt that staying healthy was an important part of their role as carer as they both felt they wanted to continue caring for their relative for as long as possible and avoid them going into a residential facility:

I suppose my biggest worry now... is my health... that I’m able to look after her. I think that worries me more than anything now – well, it would wouldn’t it - cos if I can’t do it...then somewhere down the line... what’s going to happen? ...so that worries me. So as long as I can stay reasonably fit. But no that is my main concern – and I think that a lot of people are aware of that.

(Mr Jones p.27, 1390)

And we would never wanna to put her into care or anything like that... so I just hope that obviously me and my Dad stay in good health so that we can obviously, take care of her...

(Miss Green p.32, 1518)

And finally receiving support from other people seemed important, particularly from other family members. However there was a mixed response from participants on this issue. While some reported difficulties and a distancing within family relationships, others reported experiencing good backup from theirs. For example, Mr Brown was pleased with the help that he had received from his own children in addition to his wife’s from a previous relationship:

He's the eldest... and he's been very supportive, hasn't he? He tries to come down every weekend. At one time he used to come midweek from work but of course it's such a long distance to come ... So I think he couldn't keep that up. He usually invites us up on a weekend or he comes down at the weekend. So yeah. And the other one, he comes, err, fairly regularly, our Michael, he will keep, he always phones anyway to see how you are. He's always there if you need any help anyway and, and err, his wife, err, she, Mary, she err... She takes you to have yer hair done, don't she...every month? And she's gonna take you to have yer manicure soon, come Christmas. So they're very supportive. Yeah, yeah. And my own son and his girlfriend err, they took her for a meal the other day, they're supportive as well aren't they? So, the family's being supportive.

(Mr Brown p.30, 1539)

DISCUSSION

Summary of findings

This study has explored family members' experiences in caring for someone with fvFTD. Four main themes emerged from the data. Two of these were associated with the 'emergence and realisation' of the illness; the first super-ordinate theme. In 'the opening of the eyes' there were three sub-themes, 'noticing changes', 'recounting landmark stories', and 'understanding'; which related to the participants' gradual and increasing awareness of the disease, from the initial onset and relative ignorance to increased knowledge with a greater understanding of their relative's symptoms. In the 'the double-edged sword', three further sub-themes reflected the participants' involvement with the medical and social care system: 'getting labelled', 'getting help', and 'getting researched'. What became apparent, and seemed inherent in all participants' experiences, was the two-sided nature. For instance, in order to receive benefit from the system participants had to cope with long waiting times, frustration, and uncertainty. Furthermore, while a diagnosis helps explain the changes, participants also had to face the reality of a progressive degenerative and terminal condition.

The other two main themes were subsumed under the super-ordinate theme of 'life adjustments and coping'. In 'the adaptation', three sub-themes reflected the participants' experience of becoming a carer: 'reassessing relationship', 'accepting', and 'readjusting'. It became apparent that with fvFTD the ability to relate to others deteriorates, and participants' accounts demonstrated that this had occurred early on. They described subtle changes in interpersonal relating, personality, and personal habits which altered the dynamics within the relationship. What makes this more complex is that this 'loss of relationship' begins to occur while other cognitive functions are still relatively intact; in fact participants spoke of the 'invisibility' of the condition to others. Coming to terms with the relationship changes is but one aspect, there are other uncertainties and 'strange' experiences to accept and this theme related to some compensatory intrapersonal adjustments too (e.g. developing a sense of humour). The final main theme, 'the maintenance', contained another three sub-themes all based on management (daily life, other's emotions, and self), and reflected the participants' survival strategies. All participants had taken some responsibility for caring and had developed and drawn on processes and structures in order to help them achieve this. They

also reflected on their own emotional responses to their situation and talked about how they had developed ways of coping. Moreover, they were conscious of managing others emotional experiences, from the care receiver to other family members.

‘Burden of Care’

All participants had taken on a role of caregiver (except ‘Mr Mills’ whose brother was in a specialist unit) and were potentially subject to carer stress or ‘burden of care’. They highlighted many aberrant symptoms associated with fvFTD, which indicated how they had individually perceived and interpreted them in addition to describing their management. Participants’ individuality (personality and temperament) in addition to the nature of the relationship and the availability of support seemed to subjectively affect stress levels. For example, Mrs White was managing on her own in relative isolation from family and friends with no respite care, consequently she exhibited considerable distress. Whereas Mr Brown’s distress was less evident; he had no respite care in place but was well supported by extended family members. Interestingly, Adams, McClendon, & Smyth (2008) proposed a model that places relationship factors at the centre of the stress process. They stated that the loss of intimate exchange, the change in the quality of the relationship, and an associated loss of ‘sense of self’ has a pervasive and important effect on caring for a loved one with dementia, and that this may exacerbate carer burden. The model also suggested that a process may occur during caregiving whereby a decrease in intimacy and self-identity contributes to the caregivers’ sense of role overload. This seems especially salient in this study given that the participants all reported a progressive decline in the quality of their relationships from an early stage.

Illness representations

Leventhal, Nerenz, and Steele’s (1984) self regulatory model (SRM) attempts to describe and explain how people represent and respond to health threats. It assumes that when faced with an illness people are motivated to define and control it. Central to the model is the idea that an individual actively constructs a cognitive representation of the health threat and regulates their coping in accordance with this. The self regulatory model consists of three stages: ‘representation’ or interpretation of the illness threat, ‘coping’ responses, plans, or procedures, and ‘appraisal’, the monitoring of the success or failure of those coping efforts.

The illness representation shapes the goals of coping and the use of coping strategies. There are six dimensions (Moss-Morris *et al.*, 2002):

1. Label or identity – the label of the illness, symptoms viewed as part of the disease
2. Cause – personal ideas about aetiology (simple single to more complex models)
3. Timeline / course – how long they believe it will last (acute, chronic, episodic)
4. Consequences – the expected effects and outcome of illness
5. Cure-control – how one recovers from or controls the illness.
6. Coherence – whether the symptoms seem to make an understandable whole

The SRM has been applied to understanding a wide range of chronic health conditions in recent years (Hale, Treherne, & Kitas, 2007) including dementia (Clare, Goater, & Woods, 2006; Harman & Clare, 2006), and has helped in understanding people's responses to the illness and developing suitable interventions. The SRM could therefore potentially be used to evaluate how family caregivers of people with fvFTD construe the condition, and how this relates to their coping strategies and well being. Particularly significant in the case of fvFTD, is the relative rarity of the condition and the lack of knowledge and awareness both in professional and lay circles. Carers have no reference point or information to guide their illness representations and hence their coping responses. This creates for them an extensive period of time prior to diagnosis in relative ignorance; a time in which they experience a sense of helplessness, frustration, and essentially an increased 'burden of care'.

Clinical Implications

This study highlights the need for improving dementia care services specifically to tackle the more idiosyncratic symptoms and behaviours evidenced in people with fvFTD. Firstly though, the lack of knowledge within the professional arena needs to be addressed. Access to general dementia awareness training could be increased and broadened to encompass more specific information on the signs and symptoms of fvFTD and how these differ to other dementias. This could be directed at primary care workers initially (e.g. GPs, primary care mental health workers, social workers), but could also be rolled out to care staff in residential or secondary care units. To further address the issue of uncertainty and long waiting times prior to diagnosis, clearer care pathways could be developed, reducing

frustration and distress. Furthermore, and post diagnosis, adequate follow up can be put into place by increasing provision for formal support and thereby helping to reduce isolation and increasing continuity of care. Emerging work that has developed and evaluated cognitive rehabilitation strategies indicates that these types of interventions, creatively and individually tailored, that directly target carers and their families, prove beneficial. In her modular handbook on neuropsychological rehabilitation, Clare (2008) has primarily drawn on research focussed on the primary progressive dementia associated with early Alzheimer's disease, but these principles and methods can also be applied to assist the people and their families who are experiencing other forms of dementia (and indeed other progressive neurological conditions that result in cognitive impairment). Interventions can be developed aimed at bolstering family caregivers' efficacy and self-confidence with problem-focussed coping strategies, but also bearing in mind the high emotional toll, specifically helping them to increase the use of emotion-focussed coping (e.g. acceptance). Rehabilitation approaches should therefore be holistic, taking into account both individual and systemic perspectives. In this respect support needs to be tailored for the individual and services should take into account the needs of individuals at any one time; sensitivity to the right level of intervention at the right time. Finally, mental health care policies should be designed in order to promote equality of access for all. Care should be taken to avoid ageist legislation which may unfairly limit access to effective and appropriate specialist care.

Critique

This research took a qualitative perspective using semi-structured questions in order to limit restriction placed upon participants' responses and capture the essence of their experience. IPA is an appropriate and effective methodology to achieve these aims seeking to understand how people make sense of their experiences. As with other qualitative approaches the results are subjective. However, IPA acknowledges that complete suspension of preconceptions is impossible, so the researcher made a conscious effort to be more aware of what those could be and what influence they might have on their interpretations (also see appendix 4 for a reflexive account). To take into account possible researcher effects or biases, excerpts of the transcripts and initial analyses were shared with the research and clinical supervisors to obtain feedback. This also helped in keeping the researcher focussed in a time consuming and complex analytic process. The results are not generalisable but offer a

valuable insight into a sensitive and little known subject area, grounded in the participants' own words.

Conclusions and further research

The results of this study help to shed light on the issues facing family caregivers, through presentation of an organised and coherent structure. However, it must be stressed that the intention was not to develop a linear model of 'becoming a carer'. Whilst there are some aspects that might logically follow on from others, the experience of discovering that your relative has fvFTD is less straight forward and organised. In fact it would be more fitting to consider the themes highlighted as occurring simultaneously, and dependent on individual circumstances. Further research could look at the differences between male and female family caregivers in response to fvFTD and also the experiences of adult children. More specifically studies could be designed to consider 'burden of care' issues in response to the specific symptom profile of fvFTD. It may also be valuable to examine more closely the underlying mechanism effecting the changes in relating (e.g. social cognition, empathy, and 'Theory of Mind') and to draw out the process of acceptance in family caregivers in response to this 'loss of relationship'.

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APPENDIX 1

Executive Summary

The Family Experience of Frontal-variant Frontotemporal Dementia: A Qualitative Study

This paper describes a qualitative study conducted by Paul Bradley and presented as part of a thesis for submission to the school of Psychology, University of Birmingham, for the Doctorate in Clinical Psychology.

Background and Aims of the Study

Dementia is a progressive degenerative condition caused by damage or disease in the brain. It can result in a decline in a wide range of functions controlled by the brain such as memory, concentration, language abilities or movement. Alzheimer's disease is the most well known cause of dementia commonly resulting in a gradual memory loss over time; however there are other types of dementia that have different patterns of symptoms. Frontal-variant Frontotemporal Dementia (fvFTD) can be distinguished from other dementias because it initially and primarily causes changes in behaviour and personality (with memory problems occurring much later on in the course of the disease). Dementia usually affects people in later life (over 65s) but can also occur in younger people, and fvFTD tends to develop in more people who are in their 50s-60s. The earlier onset of this condition has implications for the individual's work and family life which may be different from someone who is older. Not only will the person with fvFTD become unable to work but also family members may find fulltime employment difficult while caring for their relative. The kinds of behaviours that might occur include: shouting, swearing, and aggressive behaviour, making inappropriate comments in public, taking jokes too far, and exhibiting sexually inappropriate behaviour (Kumamoto *et al.*, 2004) and as a result caring for someone with fvFTD causes stress (Harvath, 1994). Young onset dementia is becoming more prominent now as services begin to recognise the specific needs of this younger group of people. This research aimed to address two broad questions: How does the development of fvFTD in a working age person affect the family experience of living with that person, and how might mental health services respond to the needs of those family members?

Method

Six family members of people diagnosed with fvFTD were interviewed in depth about their experiences. The interviews were semi-structured, which enabled the researcher some flexibility in choosing what questions to ask and when. It also allowed the participants the freedom to discuss their own experiences without unnecessary constraints or limitations. The interviews were transcribed and then analysed using the Interpretative Phenomenological Analysis technique (IPA; Smith, Jarman, and Osborn, 1999; Willig, 2001). IPA essentially concentrates on the meanings that people attribute to their experiences and tries to understand how they make sense of them. The researcher as part of the analytic process develops a structure of themes to demonstrate their interpretation of this.

Findings

Four main themes emerged from the data.

- In ‘the opening of the eyes’ there were three sub-themes, ‘noticing changes’, ‘recounting landmark stories’, and ‘understanding’; which all related to the participants’ gradual and increasing awareness of the disease, from the initial onset and their relative ignorance to increased knowledge with a greater understanding of their relative’s symptoms.
- In ‘the double-edged sword’, three further sub-themes reflected the participants’ involvement with the medical and social care system: ‘getting labelled’, ‘getting help’, and ‘getting researched’. What became apparent, and seemed inherent in all participants’ experiences, was a two-sided nature. For instance, in order to receive benefit from the system participants had to cope with long waiting times, frustration, and uncertainty. Furthermore, while a diagnosis helps to explain the changes, participants also had to face the reality of a progressive and terminal condition.
- In the third theme ‘the adaptation’, three sub-themes reflected the participants’ experience of becoming a carer: ‘reassessing relationship’, ‘accepting’, and ‘readjusting’. Relatively early on people with fvFTD often lose the ability to relate to others and subtle changes in interpersonal skills, personality, and personal habits change the nature of their relationships. Notably, this ‘loss of relationship’ begins to occur while other cognitive functions are still relatively intact. At the same time

relatives must also accept the ‘strange’ behaviours and might make adjustments in their own lives to compensate (e.g. develop a sense of humour).

- In the final theme, ‘the maintenance,’ all three sub-themes reflected possible management strategies for ‘daily life’, ‘other’s emotions’ and ‘self’. All participants had taken some responsibility for caring and had developed and drawn on processes and structures in order to help them. They reflected on their own difficult feelings and spoke of ways in which they had learned to cope with them, and they were also conscious of protecting the feelings of other family members as well as those of the person with fvFTD.

Discussion & Conclusions

Family members of people with fvFTD have to contend with specific behavioural challenges and personality changes associated with the condition. This requires them to learn new coping strategies and skills, while at the same time experiencing changes in the nature of their relationship, which all adds to the burden of caring. Not only that, but knowledge about fvFTD is currently limited both in professionals and the general public alike, which means that there is a prolonged period of time before diagnosis which is characterised by uncertainty and frustration; no one knows what’s wrong. After diagnosis, support services are still difficult to access because dementia services are set up to cater for older people and memory problems rather than younger people with behavioural and personality difficulties. Therefore it is necessary for health care services to be developed to cater for the specific individual needs of people with fvFTD and their families. Not only that, but fundamentally awareness about fvFTD needs to be raised in all health care services so that affected people can be supported throughout their journey from beginning to end.

References

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- Kumamoto, K., Arai, Y., Hashimoto, N., Ikeda, M., Mizuno, Y., & Washio, M. (2004). Problems family caregivers encounter in home care of patients with frontotemporal lobar degeneration. *Psychogeriatrics*, 4 (2), 33-39.

Smith, J. A., Jarman, M., & Osborn, M. (1999). Doing interpretative phenomenological analysis. In M. Murray & K. Chamberlain (Eds.) *Qualitative health psychology: Theories and methods* (p.218-240). London: Sage.

Willig, C. (2001). *Introducing qualitative research in psychology*. Buckingham: Open University.

The study is reported in detail in the following:

Bradley, P. (2009). *The Family Experience of Frontal-variant Frontotemporal Dementia: A Qualitative Study*. Clin.Psy.D. Volume I. Birmingham: School of Psychology, Birmingham.

APPENDIX 2

Literature Search Criteria

i) Databases searched

Psychinfo	Medline / Pubmed	EMBASE	Cinahl	Web of Science
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ii) Search terms and hits

SET	SEARCH TERMS USED			No. Total papers
A	Frontotemporal dementia	OR	FTD	6311
	Frontal variant frontotemporal dementia	OR	fvFTD	
	Behavioral variant frontotemporal dementia	OR	bvFTD	
	Pick's disease	OR	Dementia of frontal type	
	Frontotemporal lobar degeneration	OR	Frontal lobe degeneration	
B	Theory of Mind	OR	ToM	41075
	Social reasoning	OR	Social conduct	
	Executive function\$	OR	Executive deficit\$	
	Social cognition	OR	Social function\$	
A+B	Potential hits			366
A+B	Potential hits - Duplicates Removed			179
Total Hits (after exclusion / inclusion criteria applied)				9

APPENDIX 3

Lund and Manchester Criteria (Tables 1 & 2)

Table 1: The Clinical Diagnostic Features of FTD: Clinical Profile

Character change and disordered social conduct are the dominant features initially and throughout the disease course. Instrumental functions of perception, spatial skills, praxis, and memory are intact or relatively well preserved.

I CORE DIAGNOSTIC FEATURES

- A Insidious onset and gradual progression
- B Early decline in social interpersonal conduct
- C Early impairment in regulation of personal conduct
- D Early emotional blunting
- E Early loss of insight

II SUPPORTIVE DIAGNOSTIC FEATURES

A Behavioural disorder

- 1 Decline in personal hygiene and grooming
- 2 Mental rigidity and inflexibility
- 3 Distractibility and impersistence
- 4 Hyperorality and dietary changes
- 5 Perseverative and stereotyped behaviour
- 6 Utilisation behaviour

B Speech and language

- 1 Altered speech output
 - a. Spontaneity and economy of speech
 - b. Press of speech
- 2 Stereotypy of speech
- 3 Echolalia
- 4 Perseveration
- 5 Mutism

C Physical signs

- 1 Primitive reflexes
- 2 Incontinence
- 3 Akinesia, rigidity, and tremor
- 4 Low and labile blood pressure

D Investigations

- 1 Neuropsychology: significant impairment on frontal tests in the absence of severe amnesia, aphasia, or perceptuospatial disorder
- 2 Electroencephalography: normal on conventional EEG despite clinically evident dementia
- 3 Brain imaging (structural and/or functional): predominant frontal and/or anterior temporal abnormality

Table 2: Features Common to Clinical Syndromes of Frontotemporal Lobar Degeneration (FTLD; including: FTD, progressive non-fluent aphasia (PA), and semantic aphasia and associative agnosia (SD))

III SUPPORTIVE FEATURES

- A Onset before 65 years: positive family history of similar disorder in first degree relative
- B Bulbar palsy, muscular weakness and wasting, fasciculations (associated motor neuron disease present in a minority of patients)

IV DIAGNOSTIC EXCLUSION FEATURES

A **Historical and clinical**

- 1 Abrupt onset with ictal events
- 2 Head trauma related to onset
- 3 Early, severe amnesia
- 4 Spatial disorientation
- 5 Logoclonic, festinant speech with loss of train of thought
- 6 Myoclonus
- 7 Corticospinal weakness
- 8 Cerebellar ataxia
- 9 Choreoathetosis

B **Investigations**

- 1 Brain imaging: predominant post central structural or functional deficit; multifocal lesions on CT or MRI
- 2 Laboratory tests indicating brain involvement of metabolic or inflammatory disorder such as MS, syphilis, AIDS, and herpes simplex encephalitis

V RELATIVE DIAGNOSTIC EXCLUSION FEATURES

- A Typical history of chronic alcoholism
 - B Sustained hypertension
 - C History of vascular disease (e.g. angina, claudication)
-

APPENDIX 4

REFLEXIVE ACCOUNT

The importance of being reflexive is acknowledged within social science research and there is widespread recognition that the interpretation of qualitative data is a reflexive exercise through which meanings are made rather than found. The method and the data are not separate entities but are interdependent and interconnected and through a complex process the researcher interprets the participants' dialogue and makes choices about how and which transcript extracts to present as evidence. In this way the 'voices' of the participants do not speak on their own but are represented through the researcher. The researcher therefore unquestioningly brings to the analysis their own perspective; it is essentially a subjective interpretative process.

Firstly, I will offer some thoughts about the method and procedure chosen. My first experience of carrying out qualitative research was in 2001, where I completed a project for my undergraduate Psychology degree using Grounded Theory analysis (GT). This was a satisfying achievement although I recall complex and time consuming. I had not therefore used Interpretative Phenomenological Analysis (IPA) before and was curious to learn all about it and how it might differ from GT. I approached the project with a thirst for understanding and with a drive to learn all I could. This led to an inclination to be thorough and may have had the effect of hindering the analytic process at times, as I tended to get mired in the details, checking and rechecking categories, pondering over theme titles, and sometimes feeling overwhelmed with the mass of data being generated. However, once all the interviews had been analysed and themes were compiled and compared across transcripts, the synthesis and interpretation further refined, I felt more in control. Furthermore, at this stage the depth to which I had worked proved to be beneficial as I felt familiar with the participants' accounts before me and thankfully I had created a very clear 'paper trail' of my process. This made checking that the final themes were grounded in the participants' own words so much easier, and resulted in being able to find and highlight the most relevant quotes. Of course time constraints, word limits and the subjective nature of IPA means that the final themes presented offer just one interpretation of the data at one point in time.

I chose the subject of my research out of a genuine interest in dementia. I had worked in Older Adult psychology services before training and had an interest in neuropsychology,

conducting memory assessments in memory clinics, and also on a large national research project looking at agitation in people with Alzheimer's disease. Although most of my experience had been with Alzheimer's disease and older people I was keen to develop my knowledge on an early onset condition like frontal-variant frontotemporal dementia (fvFTD). This research project was therefore my first exposure to fvFTD.

The process of interviewing people who care for their relative with fvFTD and hearing about their lives was interesting, but challenging and also emotionally difficult at times. I was often moved when hearing about the situations that people described and the losses that they had had to deal with. I was particularly struck by the participants' difficulties in understanding the 'strangeness' of the behaviours, the dramatic changes in their relative's personality, and the distancing experienced in their relationships, and how throughout this they had all persevered. My empathising also led to my feeling frustrated when I heard time and again about the barriers that people had experienced on encountering services and in trying to understand fvFTD. I think that this offers just a little insight into the process that I underwent personally as a result of my contact with these people, and I think has a direct bearing on my interpretation of their words.

APPENDIX 5

Not available in the digital version of this thesis

APPENDIX 6

Not available in the digital version of this thesis

APPENDIX 7

Not available in the digital version of this thesis

APPENDIX 8

PARTICIPANT INFORMATION SHEET (Birmingham)

Title of Project: **THE FAMILY EXPERIENCE OF FRONTOTEMPORAL DEMENTIA**

Name of Researcher: **Mr Paul Bradley (Clinical Psychologist in Training)**

You are being asked to take part in a research study that is being carried out as part of a doctoral thesis by a Psychologist in Clinical Training. Please read the information below in order to decide if this is something that you would like to contribute to.

What is the purpose of the study?

Dementia affects people in different ways and can present them and their families with challenges in all areas of their lives. In frontotemporal dementia specific changes in the person and their lives occur and I am particularly interested in finding out how these affect and impact upon the family's daily life and wellbeing. More understanding of the issues could help shape the development of family support services in the future.

Why have I been invited?

You have been chosen because you currently have contact with clinical services in [place]. I hope to have contributions from between 6 and 10 relatives of people with frontotemporal dementia.

Do I have to take part?

You do not have to take part in the study. Neither you nor your relative's health care will be affected if you decide not to take part.

What will happen to me if I take part? What do I have to do?

If you agree to take part in the study then I will ask you firstly if you would like to take part in an interview and then at a later date a 'focus' group (an informal feedback and discussion meeting). You can choose to be interviewed and attend the 'focus' group or be interviewed only. If you agree to be interviewed then I will arrange to meet with you for between 60 and 90 minutes at a convenient location (at your home or in a clinic space) and at a convenient time. You will be asked to sign a consent form. I will then ask you about your experiences of caring for someone with frontotemporal dementia and I will record the interview on a digital recording device in order to help me to remember what you have said.

When the results of the research are completed you will be invited to attend a ‘focus’ group where we will discuss the findings of the research. Please note that the results will include direct quotes from participants. However, as part of the analysis all identifying information will be removed and participants will be given fictitious names, therefore no individual will be identifiable.

What about travel expenses and payments?

To enable you to take part I will be able to arrange transport via taxi or reimburse travel expenses, if you travel independently.

What are the possible disadvantages and risks of taking part?

This is a sensitive area and it is important to consider that, during the interview when talking about your partner or spouse’s illness, you may feel some strong and perhaps difficult emotions. If this occurs then you will not have to continue with the interview if you do not want to. We can take a break or stop completely. I will also be able to put you in touch with someone who can support you if you feel that this would be helpful.

What are the possible benefits of taking part?

Many people find it helpful to talk about their experiences and situation at length, and in detail, to an interested person. It is hoped that information that I get from this study will help shape family support services in the future.

What happens when the research study stops?

When the research study ends you will receive a summary of the results in the post. You will also be invited to the ‘focus’ group along with other interested participants.

What will happen to the results of the research study?

The research will be written up and reported in a doctoral thesis and submitted for publication in an academic journal. A public domain paper including a summary of the findings will also be sent to local NHS trusts (chief executives and directors of research). It is also possible that in the future the research will be presented at an academic conference.

What if there is a problem? How can a complaint be made?

If there is a problem then you will be able to speak directly to me or you can speak to your relative’s identified health professional at the [Service].

You will also be able to contact my academic supervisor at the University of Birmingham (please see below for contact details). If you remain unhappy and wish to complain formally, then you can contact the following person at the University of Birmingham: [name]

[name and contact details]

Will my taking part in the study be kept confidential?

Yes. I will follow ethical and legal practice and all information about you will be handled in confidence. The only people who will be aware of you taking part in the study will be me, my academic supervisor, and the clinical professional who referred you from the [Service]. All information will be kept locked in a filing cabinet or password protected on a university or my home computer. No identifying information will be included in the written transcripts or in the report and fictitious names will be assigned to each participant when the interviews are transcribed. Direct quotes from the interviews will be used in the report but no personal information will be included that will reveal your identity. The interview recordings will be destroyed after the research has been written up. Please note: If during the interview I become concerned for the current safety of you, or anyone that you speak about, then I may have to speak to my supervisor about this. If I feel that this needs to happen then I will inform you immediately during the interview.

What will happen if I don't want to carry on with the study?

If you decide at any time that you no longer wish to take part in the study then you are free to withdraw your consent, without giving a reason. If you do change your mind and decide that you do not wish to contribute then your data will be destroyed and not included in the report. Neither you nor your relative's health care will be affected if you decide not to take part.

Will my General Practitioner/Family doctor (GP) be involved?

It is not necessary for me to have contact with you or your relative's GP

Who is organising and funding the research?

The University of Birmingham is sponsoring this research

Who has reviewed the study?

All research in the NHS is looked at by an independent group of people, called a Research Ethics Committee to protect your safety, rights, wellbeing and dignity. This study has been reviewed and given favourable opinion by [Ethics Committee].

What happens next?

I will contact you by telephone to find out if you want to take part and, if so, arrange a convenient date and time to meet with you. I will also be able to arrange transport if needed.

You will be provided with a copy of this information sheet together with a signed copy of the consent form. If you have any questions or would like some more information then please telephone me on the number below.

Can I contact someone for some independent advice?

If you would like to speak to an independent professional health worker for advice about any aspect of this study then please contact:

Contact Details:

PARTICIPANT INFORMATION SHEET (Coventry)

Title of Project: THE FAMILY EXPERIENCE OF FRONTOTEMPORAL DEMENTIA

Name of Researcher: Mr Paul Bradley (Clinical Psychologist in Training)

You are being asked to take part in a research study that is being carried out as part of a doctoral thesis by a Psychologist in Clinical Training. Please read the information below in order to decide if this is something that you would like to contribute to.

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will discuss the findings of the research. Please note that the results will include direct quotes from participants. However, as part of the analysis all identifying information will be removed and participants will be given fictitious names, therefore no individual will be identifiable.

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What are the possible disadvantages and risks of taking part?

This is a sensitive area and it is important to consider that, during the interview when talking about your partner or spouse's illness, you may feel some strong and perhaps difficult emotions. If this occurs then you will not have to continue with the interview if you do not want to. We can take a break or stop completely. I will also be able to put you in touch with someone who can support you if you feel that this would be helpful.

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The research will be written up and reported in a doctoral thesis and submitted for publication in an academic journal. A public domain paper including a summary of the findings will also be sent to local NHS trusts (chief executives and directors of research). It is also possible that in the future the research will be presented at an academic conference.

What if there is a problem? How can a complaint be made?

If there is a problem then you will be able to speak directly to me or you can speak to your relative's identified health professional at the [Service]. You will also be able to contact my academic supervisor at the University of Birmingham (please see below for contact details). If you remain unhappy and wish to complain formally, then you can contact the following person at the University of Birmingham: [name and contact details]

Will my taking part in the study be kept confidential?

Yes. I will follow ethical and legal practice and all information about you will be handled in confidence. The only people who will be aware of you taking part in the study will be me, my academic supervisor, and the clinical professional who referred you from the [Service]. All information will be kept locked in a filing cabinet or password protected on a university or my home computer. No identifying information will be included in the written transcripts or in the report and fictitious names will be assigned to each participant when the interviews are transcribed. Direct quotes from the interviews will be used in the report but no personal information will be included that will reveal your identity. The interview recordings will be destroyed after the research has been written up. Please note: If during the interview I become concerned for the current safety of you, or anyone that you speak about, then I may have to speak to my supervisor about this. If I feel that this needs to happen then I will inform you immediately during the interview.

What will happen if I don't want to carry on with the study?

If you decide at any time that you no longer wish to take part in the study then you are free to withdraw your consent, without giving a reason. If you do change your mind and decide that you do not wish to contribute then your data will be destroyed and not included in the report. Neither you nor your relative's health care will be affected if you decide not to take part.

Will my General Practitioner/Family doctor (GP) be involved?

It is not necessary for me to have contact with you or your relative's GP.

Who is organising and funding the research?

The University of Birmingham is sponsoring this research.

Who has reviewed the study?

All research in the NHS is looked at by an independent group of people, called a Research Ethics Committee to protect your safety, rights, wellbeing and dignity. This study has been reviewed and given favourable opinion by [Ethics Committee].

What happens next?

I will contact you by telephone to find out if you want to take part and, if so, arrange a convenient date and time to meet with you. I will also be able to arrange transport if needed. You will be provided with a copy of this information sheet together with a signed copy of the

consent form. If you have any questions or would like some more information then please telephone me on the number below.

Can I contact someone for some independent advice?

If you would like to speak to an independent professional health worker for advice about any aspect of this study then please contact:

Contact Details:

APPENDIX 9

CONSENT FORM

Title of Project: **THE FAMILY EXPERIENCE OF FRONTOTEMPORAL DEMENTIA**

Name of Researcher: **Mr Paul Bradley (Clinical Psychologist in Training)**

Please initial boxes

1. I confirm that I have read and understand the information sheet [dated 19.09.2007; version 4 – ref: 07/Q2802/29] for the above study. I have had the opportunity to consider the information and to ask questions about the research.

2. I understand that my participation is voluntary and that I am free to withdraw at any time, without giving a reason, and without my own or my relative's medical care or legal rights being affected.

3. I agree to the interview being recorded, in order to aid the researcher's memory, and understand that when the report is written audiotapes will be destroyed.

4. I understand that information gained in the interview will be written in the report but that no individual views will be identifiable – all sources will be kept anonymous.

5. I agree to take part in the above study.

Name of Participant

Date

Signature

Researcher

Date

Signature

APPENDIX 10

The family experience of frontotemporal dementia: A qualitative exploration

Interview Guide /Schedule

Notes for the researcher:

- This document is intended to guide you through the interview.
- It lists areas / topics to discuss and a possible order [with prompts in brackets].
- It also includes a list of information at the end that if not already talked about may be useful to obtain.

NB: It is important to be flexible with its use as each individual will tell their own story in their own way.

Health & History

- What led up to you seeking help for [the patient]?
[General health, major health problems, concern about mental health?]
[Specific difficulties]
- What support do you have now?
[What support did you get or not get, what would have been useful?]

Diagnosis

- How would you describe [the patient's] condition at the moment?

[Do they have a diagnosis? When did they get it? Who gave it? Is it useful?]
[How did you feel about it? What do you think of the term frontotemporal dementia?]

Living arrangements / day-to-day life

- Can you tell me about [the patient's] and your daily life now?
[Patient living at home / care facility? since when?]
[Any care support? Respite care?]
[Info on ADLs, safety / supervising]

Effects on life

- What has changed?
[What was [the patient] like before?]
[Social / relationships – changes in, personal, other family, friends, new relationships]

[Personality – changes in, characteristics, likes and dislikes]
[Behaviours – stealing, public / socialisation, obsessions, perseveration]
- What is the most difficult aspect of life with [the patient]?
[Emotions, feelings, motivation, mood]

- How do you see your role now?
[How has it changed?]
- How do you cope?
[Coping - coming to terms with the situation/the future/getting on with life]
- How do you see your life in the future?

Check that the following has been covered during the interview...

- Ages [of the patient and interviewee]
- Relationship to the patient / how long have they known each other?
- Education & employment / occupational history
- Family – children – ages. Do they live at home? When did they leave?
Where do they live now?
How much contact do they have?
- Spouse – together, separated, divorced, widowed?

APPENDIX 11

Excerpt from transcript: Example of analytic process (extract from Miss Green)

Initial Notes	Transcript	Themes
Noticing problems at work Decreasing abilities	Pt: ..erm..you know. She was a nurse and so her colleagues noticed a change in her um, as well and they noticed that she wasn't doing her job properly.	Noticing changes (impairments at work)
Comparisons with previous abilities	R: Hmm. Pt: And then obviously they were concerned as well cos obviously is wasn't like her cos she'd been there like, 28 years.	Comparing with previous abilities
Assessing – something wrong	R: Hmm. Pt: So they knew that something was wrong, erm and she had like, tests there. R: What, cos the, the people at work had seen..?	Being assessed
Main response SHOCK – unexpected – difficult to comprehend – emotional response	Pt: Yeah. R: It had got around? Pt: Yeah. R: Mmm. Pt: So yes, yeah.. R: So she..? Pt: It's just the shock. R: Hmm. Pt: Mmm. R: Hmm. Pt: You don't expect it, do you? R: No, no. (Pause) And what does that mean for you now?	Being shocked
Loss of person – loss of relationship – loss of Mum	Pt: Um. Obviously I haven't got my Mum now. Anymore. She's here but obviously.. R: Right.	Loss of relationship
Effect of illness – does not talk	Pt: ..er, you know, she doesn't talk.. R: Hm.	Effects of illness
Echolalia – copying – odd – strange behaviour	Pt: ..a lot at all these days. She sort of copies what you say.. R: Right. Pt: ..so it just means not having her there to talk to and.. R: Yeh.	
Strangeness – difficult to comprehend	Pt: ..you know.. R: Mm hm. Pt: ..it feels like she's not here, because..she's only here in...	

Excerpt from transcript: Example of analytic process (extract from Mr Mills)

Initial Notes	Transcript	Emergent Themes
Protecting other peoples' emotions Taking on responsibility	Pt: I protect them really as much as I can because: they don't need to know the ins and outs of what John's doing.. R: Mm. Mm. Pt: Um. Yeah. I try to protect them as much as I can. R: And I guess it's difficult. You can't really be definite about things really, can you, cos it's..? Pt: Not at all. No. It's an unknown quantity for me as well.	Protecting others Managing others
Unknown quantity Vague Unnerving uncertainty	R: M hm. Pt: You know..it.. (Pause) I won't say it's difficult, but it's unnerving at times, definitely. R: Unnerving? Pt: Yeah. Yeah. R: How do you cope with that? Pt: (Short pause) Arhm. I think because he's so far away.., perhaps I only see him every two weeks, as an average..	Recognising own emotions
Having distance – not seeing him too often Separated parts of live	R: Mm. Pt: it's like er.., I don't know, it's.. I come down, I deal with it and go away. I don't dwell on it. The only times I dwell on it is if my Mum comes down, Mum and Dad come down and there's been a problem.	Coping – compartmentalising Distancing in relationship
Not dwelling on things – getting on with it	R: Mm. Pt: Arhm. For instance, he didn't go out: I'm thinking, "why has he done that? Is that a deterioration?" So I perhaps ring the Unit manager the following week to find out...	Not dwelling on it Managing self
Taking action – taking control – doing – coping – problem solving	R: What he's doing, that kind of thing? Pt: ..find out what it's all about and she'll say: "Well actually he's been unwell for the past couple of days." Whatever. And because my Mum and Dad come down and they	Managing – problem solving Taking control

APPENDIX 12

Example of a range of themes from one transcript
Mr Mills – Final Themes

EMERGENCE & REALISATION

Becoming aware / onset

- Initial problems (noticing changes, something amiss, putting the pieces together)
- Relationship difficulties

Landmark events / turning points/ important stories

Effects of Illness – the symptoms

- Explanations / trying to make sense
- Understanding John's problems
- Dealing with challenging / difficult behaviours
- Dealing with change / deterioration – progression / changing course

Comparisons with past - past functioning

Learning about the disease & taking action (to learn & get treatment), understanding the disease

Entering the system

- Accessing services
 - Quick service
 - Diagnosis
 - Waiting
 - Barriers to access
 - Lack of specialist knowledge / understanding (professionals)
 - Lack of specialist services
 - Challenging/negotiating with the system
 - (Dealing with bureaucracy, being assertive, taking control, compromising)
 - Being supported by the system
- Being part of a clinical trial

LIFE ADJUSTMENT & COPING

Coping with own emotions

- Recognising them, distancing from them (having a clinical / detached approach / using humour)

Not dwelling on it (think - take action)

Carer stress / burden

Living with it

Living with uncertainty (apprehension, what comes next, vagueness)
Coming to come to terms with it (trying to understand, being realistic, wondering about the future; drugs)

Loss of relationship

Rejecting him
Loss of control

Managing him

Problem solving
Being exposed to the public
Keeping him safe
Supervising him
Keeping routine / normality
Manipulating him

Managing other people

Taking responsibility / a new role
Managing other people's emotions (manipulating the facts / watering down)
Protecting the children (kid language v isolating)