

Specific support needs and experiences of carers of people with frontotemporal

dementia:

A systematic review

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Abstract

Introduction: Frontotemporal dementia (FTD) is one of the most common types of dementia in persons younger than 65 years of age. Diagnosis is often delayed due to slow gradual decline and misinterpretation of ‘non-typical’ dementia symptoms. Informal carers of people with FTD experience greater levels of overall burden than carers of people with other forms of dementia. Aim of this systematic review was to describe the subjective experience of being an informal carer of a person with FTD, and to identify the specific needs, coping strategies and helpful support resources of this carer population.

Methods: Four electronic databases were used to search for published literature presenting experiences of carers of people with FTD between January 2003 and July 2019. Search strategy followed PRISMA guidelines. Findings were analysed using Framework Analysis, employing five stages of analysis to develop a coding index and thematic framework that included key aspects of the carer experience, which were grouped into themes and presented in a narrative format.

Results: 1213 articles were identified in total. Twelve studies were included in the final synthesis of the review. Six themes were identified: ‘Challenging road to and receipt of diagnosis’, ‘relationship change and loss’, ‘challenging experiences in caring’, ‘positive experiences and resilience’, ‘coping, and support needs’.

Discussion: Findings highlight an increased need for carers of people with FTD to receive support during the pre-diagnostic stage, including support to manage symptoms. Further research should explore relationship changes and loss amongst carers to inform approaches for carer support. In conclusion, the lack of knowledge and unique needs of carers highlight the importance of public awareness campaigns and healthcare professional education to support carers with FTD symptom impact.

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Keywords: Frontotemporal Dementia, Frontal temporal dementia, carer, caregiver, carer support, spousal carer, carer coping

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Introduction

Frontotemporal dementia (FTD) is a form of dementia that affects mainly the frontal and/or temporal lobes in the brain (Bang et al., 2015). FTD is a heritable neurodegenerative disorder, with approximately 30 per cent of patients having a familial (genetic) variant of FTD (fFTD; Seelaar et al., 2011), classified by having a family history of FTD and an inherited mutation (AFTD, 2012; Seelaar et al., 2011). Although there has been an increase in research in the past decade to understand FTD pathology, genetic links, and prognosis, research is required to understand the needs of families of people living with these diseases (Hodges & Piguet, 2018). Recent reports suggest that while FTD accounts for less than 20 per cent of all dementia diagnoses (Leroy et al., 2021; Religa et al. 2015; Van der Flier and Sheltens, 2018), it is a common cause of young-onset dementia for people under the age of 65 (Coyle-Gilchrist et al., 2016; Karageorgiou and Miller, 2014; Rosness et al., 2016). Despite a varied age of onset, the mean age at diagnosis is 58 (Leroy et al., 2021). However figures reported should be considered with caution given that FTD remains under-diagnosed (Beach et al., 2015) misdiagnosed (Berber and Chaves, 2013; Giamarelou et al., 2017)), and often time to diagnosis is longer than for other dementias (van de Vliet et al., 2013), suggesting delays to diagnosing FTD. Additionally, with some exceptions (e.g. Onyike & Diehl-Schmid, 2013; Leroy et al., 2021), many estimations have been made using previous diagnostic criteria (i.e. Lund and Manchester groups, 1994; Neary et al., 1998), despite changes being made to diagnostic criteria in the early 2000s (i.e. Hodges & Patterson, 2007; Gorno-Tempini et al., 2011; Rascovsky et al., 2011; Rohrer et al., 2010).

Despite the importance of an early diagnosis, this complex presentation of symptoms can often extend time to diagnosis to up to 5 years from the point of noticing initial changes (Rosness et al. 2016). FTD is marked by a gradual onset (Johannessen et al., 2017). Often family members may therefore misinterpret subtle symptoms as variations of a person's

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mood and personality, often putting them down to fatigue, stress, overwork, or depression (Ducharme et al., 2013). Attributing symptoms to disease may be further complicated by the person with FTD, who often lacks insight (Ducharme, et al., 2013).

On average people live for about six and a half years after family members notice the initial symptoms (Xie et al., 2008), and whilst there is no cure for FTD (Rosness et al., 2016; Warren et al., 2013; Young et al., 2018), treatments currently used include symptomatic drug therapies such as selective serotonin reuptake inhibitors (SSRIs), and psychosocial interventions offered to support patients, families and carers to manage and cope.

People with FTD are less likely to present initially with symptoms of memory loss or visuospatial impairment (Warren et al., 2013). Neuronal loss in the frontal and temporal brain regions of people with FTD is responsible for behaviour, executive functioning, and language, and damage to this area impacts personality, emotional control, planning, problem-solving, and speech (Warren et al., 2013). FTD is often divided into a behavioural variant (bvFTD) and a language variant (known as primary progressive aphasia; Mioshi et al., 2009; Warren et al., 2013), although these commonly overlap, and may also be accompanied by neuropsychiatric symptoms, including anxiety, depression, and psychosis (Rosness et al., 2016).

Many people with FTD are cared for at home by family members or spouses, and are referred to as ‘carers’, ‘family carers’ or ‘informal carers’, with these terms often used interchangeably in the literature. The World Health Organisation (WHO) defines carers by the relationship (including spouses, children, daughters and sons-in-law, friends, neighbours), by living arrangements (including those residing with the care recipient or living separately) and the level of care input (regular caring, occasional or routine; WHO, 2012). Carers can be involved in providing direct care and/or indirectly, for example by organising care delivered by others (WHO, 2012).

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Recent reviews of the literature have reported that being an informal carer for a person with dementia can lead to physical and psychological problems, social isolation and financial difficulties (Brodaty & Donkin, 2009; Chiao et al., 2015; Lindeza et al., 2020; Lindt et al., 2020). These areas of difficulty are often combined in the measurement of carer ‘burden’, which refers to the physical, financial and emotional strain experienced by a carer of a person with illness (Dunkin & Anderson-Hanley, 1998). Evidence from quantitative research suggests that carers of people with FTD experience greater overall burden than carers of people with other forms of dementia, such as AD (de Vugt et al., 2006; Kaiser & Panegyres, 2007; Riedijk et al., 2006). In a study conducted in the Netherlands, authors suggested that increased carer burden is largely associated with the behavioural changes of FTD (De Vugt et al., 2006). Subjective reporting of FTD carers’ experience has been limited, and has not explored areas of resilience or positive aspects of caring reported in general dementia carer literature, such as an increased family cohesion and a sense of personal achievement, growth and purpose in life (Yu, Cheng, & Wang, 2018). The existing qualitative studies in the topic area, have frequently combined FTD with other forms of dementia or early onset dementia in the reporting of results (Ducharme et al., 2014, 2013), making it difficult to distinguish between the experiences of carers of people with FTD and those of people with other forms of dementia.

While there have been syntheses of burden literature for carers of people with FTD, to our knowledge there has not been a systematic synthesis of the support needs and subjective experiences of carers of people with FTD. This has been due in part to the primary focus on burden, and the methodologies of existing reviews, which have included studies not limited to FTD. Therefore, existing reviews do not clearly depict the specific needs and experiences of FTD carers.

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The objectives of the review include: a) To integrate the findings from research examining subjective experience of being an informal carer of a person with FTD, b) to identify the specific needs of informal carers of people with FTD, and c) to identify coping strategies and helpful support resources used by informal carers of people with FTD.

Method

Overview

This review was conducted using the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) Checklist (Moher et al., 2009). The review protocol was registered on the International Prospective Register of Systematic Reviews—CRD42019149625.

Literature Search

The search strategy included free text and expanded subject (MeSH) terms, carried out using four electronic databases: PubMed, PsycINFO, Current Nursing and Allied Health Literature (CINAHL), and Thomson Reuters Web of Science electronic databases to identify articles from 2003 (due to the advent of the diagnostic criteria used in research at this time) to Week 3, July 2019. Additionally, reference lists were reviewed from primary studies and systematic reviews, and Google Scholar and Web of Science citation searches were conducted. Authors of included studies were contacted to request additional details, where important information was unclear or missing for the purpose of the review.

The search was conducted with the following search terms : a) Caregiver, carer *and* b) Frontotemporal dementia, *and* c) Needs, coping, experience, support. See Appendix A for complete list of search terms. Duplicates were excluded.

Inclusion, Exclusion Criteria and Data Collection

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Inclusion criteria were: a) Quantitative, qualitative, and/or mixed method studies; b) non-interventional studies; c) Reported results relating to needs, experience or coping strategies of informal carers of adults with FTD; d) Sample were informal family carers of adults with FTD; e) English language or translation available articles; f) Peer reviewed; g) Published between June 2003 and June 2019. The review applied the WHO (2015) definition of ‘informal carers’, as stated in the introduction section.

Exclusion criteria were: a) Pooled samples of carers of FTD with other dementias; b) no reported diagnosis of FTD; c) case series or case report, review studies, letters to the editors, professional opinions, dissertations or conference abstracts; e) reported only carer burden; e) reported only comparison findings between FTD and other dementias; f) focus only on patient (not carer).

The first author independently screened each study by title and abstract. Full-text was acquired for all included articles and reviewed against the inclusion and exclusion criteria. Included articles were reviewed by a second researcher (*insert initials of author*) and disagreements resolved through discussion.

Quality Assessment

The first author independently assessed risk of bias using the Mixed Methods Assessment Tool (MMAT; Hong et al., 2018), which has been designed to assess for quality in systematic reviews that include quantitative, qualitative and mixed-methods studies. Authors of MMAT (Hong et al., 2018) discouraged calculation of an overall score, therefore each criterion was rated for quality using broad categories of ‘low’, ‘medium’, ‘good’, and ‘high’, according to the criterion.

Strategy for data synthesis

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Quality scores were used to remove poor quality studies (those of 'low' quality excluded prior to synthesis), to weight findings according to the methodological rigour and inform interpretation of competing findings (findings from higher quality studies prioritised). Aggregate data were presented in a narrative synthesis, aiming to describe the experiences and support needs of carers of people with FTD, as represented in existing literature. Qualitative themes were synthesized using a framework analysis (Ritchie and Spencer, 1994) to develop themes, presented in a narrative format. The recommended five stages of analysis (familiarisation, identifying thematic framework, indexing, charting, mapping and interpretation; Ritchie and Spencer, 1994) were used to develop a thematic framework and coding index that included key aspects of the carer experience. This included positive and negative experiences, carer support needs, and coping strategies used amongst carers of people with FTD. Quantitative findings were then incorporated within the corresponding themes delineated from qualitative synthesis and reported within these themes in the write-up of the results.

Results

Searches produced 1213 articles, reduced to 513 following exclusion of duplicates. Screens of title and abstract removed 480 articles. Inclusion and exclusion criteria were used in the full-text review of thirty-three articles. Twenty-one articles were excluded during this process, resulting in twelve included studies. Twelve studies were then included in the data extraction and final synthesis of the review. See Figure 1 for details. See Appendix B for complete report of articles excluded at the full text review stage.

Study and Participant Characteristics

Methodology. Of the twelve studies included in the review (participant $n = 549$), five employed a qualitative methodology (Johannessen et al., 2017; Massimo et al., 2013; Nichols, et al., 2013; Oyebode et al., 2013; Rasmussen et al., 2019) and seven were

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quantitative (Chow et al., 2011; Diehl-Schmid, et al., 2013; Nicolaou et al., 2010; Riedijk et al., 2008; Riedijk, 2009; Roche et al, 2015; Wong and Wallhagen, 2014). Two of the included studies were from Canada, two from the USA, two from Australia, two from the Netherlands, and four from elsewhere in Western Europe (UK, Germany, Norway and Sweden).

The most common quantitative measures of carer experience were the Neuropsychiatric Inventory (NPI; Cummings et al., 1994) (3 studies), the Health related Quality of Life 36-item Short Form Health Survey (SF-36; Ware & Snow, 1994) (3 studies), and various measures of mood including the Revised Symptom Checklist (SCL-90-R; Derogatis and Cleary, 1977) (2 studies), and the Beck Depression Inventory, second edition (BDI-II; Beck, Steer, and Brown, 1996) (2 studies). These included the Social Support List (Van Sonderen, 1993), the Resource Utilization in Dementia Lite (RUD-lite; Wimo et al., 2013), and a non-validated measure, which reviewed support services and interventions, with ratings on helpfulness and need (Diehl-Schmid et al., 2013). Other factors of the carer experience were measured, including coping strategies employed by carers, which were measured using the Brief COPE Scale which is a 28-item self-report questionnaire (Carver, 1997) (2 studies), and one study employed the Utrecht Coping List (Schreurs, 1993). Three studies employed quantitative measures to identify support needs of carers, however there was no consistency in measures of patient carer relationship and carer needs used. For a full description of studies see Table 1.

Table 1*Description of Studies*

Reference	Database	Study aims	Study design & Sample (N=549)	Measures	Main findings	Key limitations
Chow et al., 2011 [Canada]	PsycINFO CINAHL	Compare impressions of FTD carers & track care-recipient symptoms to inform support interventions for carers.	Quantitative Cross-sectional survey (n=79)	Survey & tracking instrument provided by Dementia Guide, Inc., (http://www/dementiaguide.com)	<i>Carer experience:</i> 50% carers reported greatest difficulty during pre-diagnostic stage due to a) behavioural and personality changes not identified as symptoms of disease, and b) delayed diagnosis. c) 24% of sample lacked confidence in carer role, d) 20% discovered strength in caring role. <i>Support:</i> Family counselling and the public education about young onset dementia.	a) Recruitment bias: Excludes those without internet access. Snowball recruitment- carers recruited in clinic.
Diehl-Schmid et al., 2013 [Germany]	PsycINFO	Identify problems and needs of FTD carers, examine factors associated with carer strain & depression, & determine helpfulness of support interventions.	Mixed-Methods Cross-sectional questionnaire (n= 94)	<i>Carer Experience:</i> Negative impacts of caring, carer-patient relationship using Resource Utilization in Dementia lite (RUD-Lite) (Wimo & Winblad, 2003), Beck Depression Inventory (BDI-II; Beck et al.,1996). <i>Carer Support:</i> List measuring 45 support services & interventions.	<i>Carer Experience:</i> Care-recipient's behaviour and in the interpersonal relations between carers and care-recipients were associated with carer depression. <i>Support:</i> a) Need information regarding FTD to be provided to family and professionals, b) psychosocial support through educated staff, c) financial support.	a) Self-reported diagnosis by carers. b) Unclear mixed methodology. c) may not be generalizable outside Germany.
Johannesen et al., 2017 [Norway]	Web of Science	Examine the spouses of yoFTLD experiences and	Qualitative Corbin and Strauss (2008) modified	In-depth Interviews & a thematic analysis	<i>Carer Experience:</i> 1) 'Sneaking signs at the early stage of dementia'- incomprehensible early signs and lack of self-insight. 2) 'Other relations'- the torment, interference with work and vanishing social relations.	a) Small sample size.

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		needs for daily assistance.	Grounded Theory (n=16)		<i>Support:</i> 'Needs for assistance through all stages of dementia'	
Massimo et al., 2013 [USA]	PsycINFO & Pubmed	Explore lived experience of caring for a person with bvFTD from the perspective of two spousal carers.	Qualitative Brenner's Interpretive Phenomenological (Brenner, 1994) (n=2)	In-depth interview & a thematic analysis	<i>Carer experience:</i> 1) Identity and role change- Loss of previous identity, of marital connections, of shared 'taken for granted' social meanings and shared meaningful future; 2) Isolation, due to care-recipient lacking insight; 3) Anger, due to care-recipient erratic behaviour, emotional coldness and lack of reciprocated feelings by the care-recipient; 4) Facing the future and building ideas for a good day. <i>Coping:</i> Reminding self that pathology of FTD causes the behaviour difficulties.	a) Small sample; b) Data collection methods may not be fully in line with methodology (e.g. Closed questioning employed)
Nichols et al., 2013 [Canada]	PsycINFO	Identify needs & experiences of young carers of people with FTD, to inform the development of a support website for young carers.	Qualitative Thematic Analysis (Braun and Clarke, 2006) (n=14)	Focus groups & a thematic analysis	<i>Carer experience:</i> 1) Emotionally impacting experience, 2) caregiving experience, rewards and challenges 3) difficult experiences as a result of personality and behavioural changes, 4) child-care-recipient relationship changes. <i>Support:</i> 1) Improved pathway to diagnosis. 2) Education about FTD. 3) In-person support from family, friends and professionals. 4) age-stratified support groups. <i>Coping:</i> 1) Maintaining positive but realistic view, 2) have compassion for the person with FTD.	a) Possible bias in data collection procedure: focus group leader was parent to two young carer participants.

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Nicolaou et al., 2010 [Australia]	Backwards search	Explore needs, level of burden, depression and anxiety of FTD carers and AD carers.	Qualitative Cross-sectional questionnaire (n= 60)	<p><i>Carer Experience:</i> Depression, Anxiety and Stress Scale (DASS; Lovibond & Lovibond, 1995) to measure depression, anxiety and stress. <i>Carer needs:</i> Camberwell Assessment of Need for the Elderly (CANE; Orrell & Hancock, 2004; Reynolds et al., 2000) with two carer-related areas (i.e., information and psychological distress).</p>	<p><i>Carer needs:</i> All carers reported at least 12 needs, with at least 6 unmet needs (greater than AD). Increased need related to the younger onset of FTD, financial dissatisfaction, typical FTD characteristics, access to appropriate services, information and support.</p> <p><i>Carer Experience:</i> Difficulties with household activities, food, self-care, daytime activities, communication, continence, psychological symptoms, information, deliberate and accidental self-harm, abuse, behaviours, social company, money, information for carers and carer distress were significantly greater, and viewed primarily as unmet needs by FTD carers, with greater need than AD carers.</p> <p><i>Support:</i> 1) Specific educational & support programs to increase community awareness of FTD, adapt residential services & activities for people with FTD.</p>	a) Unclear what services people were receiving at time of data collection.
Oyebode et al., 2013 [UK]	PsycINFO & Pubmed	Explore experiences of having a relative with bvFTD, to inform services to meet family carers' needs.	Qualitative Interpretative phenomenological analysis (IPA; Smith et al., 2009) (n= 6)	Semi-structured interview & a thematic analysis	<p><i>Carer Experience:</i> 1) Witnessing bizarre and strange behavioural and personality changes, 2) Managing behaviours - includes defending & explaining behaviour, promoting quality of life, and working around lack of insight; 3) Impact on the relationship and the person with FTD - includes loss of the person and relationship, grief.</p> <p><i>Support:</i> 1) Services to reduce stigma and improve awareness of FTD and support for care-recipients. 2) Sharing internal and</p>	a) Findings may not be transferable to target population, due to mixed sample. c) Missing definition of "emotional involvement and decision making" of participants, creating unclear overlap with carer role.

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Rasmussen et al., 2019 [Sweden]	PsycINFO, CINAHL & Pubmed	Explore family carers experiences of the pre-diagnostic stage of FTD	Qualitative Gadamerian hermeneutic method (Gadamer, 1999; Fleming et al., 2003) (n=14)	Semi-structured interview & a thematic analysis	<p>external resources for support & support from family & friends.</p> <p><i>Coping:</i> Personal hobbies and interests, focusing on the present, blaming the illness not the relative, and reminding oneself that matters could be worse.</p> <p><i>Carer Experience:</i> 1) 'Becoming distant' - silence and apathy creating distance in the relationships. 2) Changes misinterpreted as due to natural aging, somatic or psychiatric illness; 3) 'Becoming insecure' - due to vague symptoms, experienced as strange, frightening, irritating, & difficult to explain to others; 4) 'Becoming devastated' - Feelings of guilt, fear and anger regarding dangerous or risky behaviours, led to catastrophic thoughts and insomnia; 4) 'Becoming a stranger' - loved one became a stranger due to changes in behaviour and personality, and the care-recipient's lack of insight regarding these changes. 5) Carer took on a different role in the relationship. Experienced guilt, exhaustion and sometimes depression.</p> <p><i>Support:</i> 1) Need for increased awareness of FTD amongst clinicians and specialists; 2) Need for clinicians respond to families' concerns about personality & behavioural changes & loss of function.</p>	a) Interpretation is only one of several possible ones.
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Riedijk et al., 2008 [The Netherlands]	PsycINFO & CINAHL	Understand change in carer burden & relationship quality between informal carer and person with FTD over 2 year period, from baseline.	Quantitative Longitudinal questionnaire study (n=63)	<p><i>Carer Experience:</i> Neuro-psychiatric Inventory (NPI; Cumming et al., 1994)-measures emotional difficulty due to care-recipient neuro-disturbance. Measures carer general psychopathology with Health related quality of life (HQoL SF-36; Ware & Snow, 1994). Relationship quality measured using 4 items of the USC Longitudinal Study of Three-Generation Families measures of positive affect (Lawrence et al., 1998).</p> <p><i>Coping & Support:</i> Utrecht Coping List (Schreurs, 1993). The Social Support List (Van Sonderen, 1993).</p>	<p><i>Carer Experience:</i> 1) Psychological well-being remained stable over a 2 year period. Carers of people with FTD living in the home (FTDH) experienced greater difficulties than carers of people with FTD living in nursing homes. 2) Relationship closeness preserved, whereas communication and sharing viewpoint on life significantly reduced.</p> <p><i>Coping and Support:</i> 1) All coping strategies were used to a similar degree from baseline to 24 months except for depressive reaction, which increased from baseline to 24 months ($p > 0.05$). 2) Emotional expression was used more ($p > 0.001$) by FTDN carers than FTDH carers.</p>	1) Small sample size, potentially affecting detection of significant differences. 2) Potentially biased sample due to high drop out rate, (24 out of 63).
Riedijk et al., 2009 [The Netherlands]	PsycINFO & CINAHL	Assess degree to which FTD carers felt competent in caregiving, to evaluate the relationship between competence and carer burden. Timepoint: at 24 month follow-up	Quantitative Cross-sectional questionnaire (n= 46)	<p><i>Carer experience:</i> Sense of Competence Questionnaire (Vernooij-Dassen et al., 1996); NPI (Cumming et al., 1994); SCL-90-R (Derogatis & Cleary, 1977); HQoL SF-36 (Ware & Snow, 1994).</p>	<p><i>Carer Experience:</i> 1) Carer self-confidence was not related to dementia severity. 2) Competence was associated with emotions & attributions of care-recipient's behaviour and carer sacrifice. 3) Sacrificing one's personal life to caring was associated with more psychological complaints & reduced quality of life.</p> <p><i>Support:</i> Authors suggest carers may benefit from psycho-education on self-care and methods to allow carers to have time to meet their own needs. Model developed to explain this.</p>	Same as above

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Roche et al., 2015 [Australia]	PsycINFO	Identify coping strategies that influence psychological outcomes of carers of people with FTD, assess degree that carers felt 'adequate' in their caring role, and to evaluate the relationship between carer self-confidence & burden.	Quantitative Cross-sectional questionnaire (n= 94)	<i>Carer Experience:</i> BDI-II (Beck, Steer, & Brown, 1996); Quality of Life-Alzheimer's Disease (QoL-AD; Logsdon, Gibbons, McCurry, & Teri, 1999). <i>Coping & Support:</i> The Brief COPE Scale (Carver, 1997) to assess the participants' coping strategies .	<i>Carer Experience:</i> 1) The effect the care-recipients' care needs on carer well-being depended on carers' experience of strain. High levels of carer strain predicted reduced QoL (-0.44, CI: [-1.15 to -.16]), but not depression (-0.22, 95% confidence interval CI: [0.16 to 2.04]). 2) Carers' experience of strain was exacerbated by their use of dysfunctional coping (beta = .21; p = .04), care-recipients' intensity of care needs (beta = .25; p = .01), and fewer financial resources (beta = .23; p = .02). <i>Coping:</i> 1) Carers' use of dysfunctional coping as a response to their strain increased the levels of depression (0.46, CI: [0.19-0.82]). 2) Use of problem-focused coping strategies increased carers' QoL (0.10, CI: [0.00 to 0.31]).	a) Possible recruitment bias, via FTD support groups and associated organizations; b) Study design did not permit individualised standardised testing & rating of dementia severity for care-recipients; c) Measures do not directly address subjective appraisals of caring.
Wong & Wallhagen, 2014 [USA]	CINAHL	Examine the relationship between coping and carer physical & mental health among family carers of people with FTD.	Quantitative Cross-sectional questionnaire (n= 61)	The Brief COPE Scale (Carver, 1997); Couple's Satisfaction Index - 4 (CSI-4; Funk & Rogge, 2007); NPI; Cumming et al., 1994; HQoL SF-36 (Ware & Snow, 1994)	<i>Carer experience:</i> 1) A small positive association was identified between problem-focused coping and carer physical health (r = 0.29, p < 0.05). 2) Emotion-focused coping ($\beta = 0.46$, p < 0.05) made a statistically significant, unique contribution to carer mental health and explained approximately 14% of its variance.	a) Low internal consistency for two coping subscales; b) Diagnosis of FTD based on carer self-report; c) possible recruitment bias via membership to FTD support group and associated organizations.

Participant Characteristics. Two studies shared a database and the same cohort (Diehl-Schmid et al., 2013; Roche et al., 2015), and two studies were from a longitudinal study at two different time points: data was collected at baseline, 6, 12 and 18 months (Riedijk et al., 2008) and at 24 month follow-up (Riedijk et al., 2009). Sixty-seven per cent of carers were female ($n = 368$), with an average age of 65.52. The majority of carers were partners or spouses and one study sample consisted only of children of people with FTD (mean age 14.28, range 11-18; Nichols et al., 2013). Only four studies reported carers with child dependents, including the child participant study ($n = 39$; Diehl-Schmidt et al., 2013; Johannessen et al., 2017; Nichols et al., 2013; Roche, et al., 2015). Care-recipients had a diagnosis of FTD, with the majority reporting with behavioural symptoms indicative of bvFTD, with the exception of the Chow, Pio and Rockwood (2011) study, which was reported to have a higher response rate from carers of people with PPA, resulting in a sample that may be more representative of carers of people with largely language-based difficulties. One study (Nicolaou et al., 2010) reported a mixed sample including care recipients with AD, however only data from results reported for carers of people with FTD was extracted. The majority of the care-recipients were male. The mean time to diagnosis was about three and a half years (range = 0-12 years). Four studies reported the age at time of diagnosis, with the mean age at approximately 57 years of age (range = 28-88). Ethnic and cultural descriptors were not reported. For full description of participant characteristics see Appendix D.

Quality of included studies. Six studies were rated as of 'Good' quality (Diehl-Schmid et al., 2013; Nichols et al., 2013; Riedijk et al., 2008; Riedijk et al., 2009; Roche et al., 2015; Wong and Wallhagen, 2014), four were 'High' (Johannessen et al., 2017; Nicolaou et al., 2010; Oyeboode et al., 2013; Rasmussen et al., 2019), two were of 'Medium' quality (Chow al., 2011; Massimo, Evans & Benner, 2013), and none received a low rating.

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Therefore, no article was removed due to low methodological rigour. See Appendix C for synthesis of quality assessment results.

Methodological issues. Most methodological issues were related to possible sampling bias, whereby the recruitment methods may have contributed to a non-representative sample, given that many participants were recruited via snowball methodology (Chow, Pio & Rockwood, 2011; Wong and Wallhagen, 2014), clinician identification of participants (Massimo et al., 2013; Oyeboode et al., 2013; Johannessen et al., 2017; Rasmussen et al., 2019) or by participating in an FTD support group (Diehl-Schmidt et al., 2013; Wong and Wallhagen, 2014). Furthermore, sampling bias may have been present in four studies, which relied on carer self-report as a caregiver of a person with FTD, without systematic measurement or review of medical notes to confirm diagnosis (Chow et al., 2011; Diehl-Schmidt et al., 2013; Nichols et al., 2013; Wong and Wallhagen, 2014). Two longitudinal studies had high dropout rates, which may have biased samples (Riedijk et al., 2008; Riedijk et al., 2009). One study presented possible bias in data collection procedure, whereby the focus group facilitator was a parent to two teenagers in a family impacted by FTD, and these two children participated in focus groups (Nichols et al., 2013).

Narrative Synthesis

Framework analysis was applied as detailed in the methods, and produced six themes. Themes depict carers' experience of: a) the challenging road to and receipt of diagnosis, b) relationship change and loss, c) challenging experiences in caring, d) positive experiences and resilience, e) coping, and f) support needs.

Challenging road to and receipt of diagnosis. Carers experienced a challenging road to receiving a diagnosis of FTD, including the pre-diagnostic experience of carers (Chow et al., 2011; Johannessen et al., 2017; Nichols et al., 2013; Oyeboode et al., 2013; Rasmussen et al., 2019). The road to diagnosis included the carers' difficulty with initial

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symptom recognition and attribution, whereby carers had difficulty identifying symptoms, and explaining them to others, due to the often subtle and gradual behavioural and personality changes. During the pre-diagnostic stage, carers often misinterpreted symptoms, believing that they were signs of physical or psychiatric illness, the natural course of aging (Rasmussen et al., 2019), or variations of their normal personality (Johannessen et al., 2017), rather than signs of dementia. This difficulty identifying symptoms was reported to have a negative impact on carers' wellbeing. Qualitative studies described how carers experienced these initial changes and reported them to be strange, frightening to witness, irritating, and socially embarrassing (Johannessen et al., 2017; Massimo et al., 2013; Nichols et al., 2013; Oyeboode et al., 2013; Rasmussen et al., 2019).

Carer's described the experience of receiving a diagnosis as with shocking and confusing (Nichols et al., 2013; Oyeboode et al., 2013), and although carers described this as a difficult experience, one study reported that diagnosis came as a relief to some, reporting that it provided the opportunity to receive support from services (Johannessen et al., 2017). Nevertheless, some were unable to access support due to the person with FTD lacking insight and being unable to accept their diagnosis, placing continued burden on carers (Johannessen et al., 2017; Massimo et al., 2013; Oyeboode et al., 2013). Studies reporting these findings were qualitative in methodology.

Relationship change and loss. Carer's experience challenges caused by the symptoms of FTD, which dramatically changed the relationship dyad to such an extent that carers came to change their view of themselves. Studies reported associations between this shift and a growing distance in the relationship, leading the carer to grieve the loss of the person with FTD, their mutual relationship, and the loss of one's previous identity as a spouse or child, which was gradually replaced by the carer role (Diehl-Schmid et al., 2013; Massimo et al., 2013; Oyeboode et al., 2013; Rasmussen et al., 2019).

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Carer's experience negative aspects when taking on the carer role, which include an increasing sense of isolation, as the care-recipient becomes more dependent, their insight and apathy toward them increases and communication deteriorates, making meaningful interactions with the care-recipient challenging (Diehl-Schmid et al., 2013; Johannessen et al., 2017; Massimo et al., 2013; Nichols et al., 2013; Oyeboode et al., 2013; Riedijk et al., 2008; Wong and Wallhagen, 2014), as reported in both quantitative and qualitative findings. Quantitative findings from a mixed-methods study identified that a worsening carer-patient relationship and the younger age of the patient at diagnosis was significantly associated with carer depression ($p = 0.048, 0.001$; Diehl-Schmid et al., 2013). Surprisingly, another quantitative study reported that 'relationship closeness' and 'getting along' in the carer-patient relationship was preserved over time (Riedijk et al., 2008). Authors of this study suggested that a high quality of 'pre-morbid' relationship might act to protect carers against the negative impacts of caring, despite increasing severity of FTD (Riedijk et al., 2008).

The carer role includes typical caring duties (e.g. coordinating medical and social services, delivering care, taking household and child-care responsibilities, etc.). Specific experiences related to FTD symptoms includes managing behavioural changes in the social realm and coping with feelings of isolation and social embarrassment regarding behavioural symptoms (Nichols et al., 2013; Oyeboode et al., 2013; Rasmussen et al., 2019), as well as educating others about FTD (Nichols et al., 2013; Oyeboode et al., 2013) and were depicted in qualitative studies.

Challenging experiences in caring. Carers experience challenges and difficult emotions when witnessing and/or managing care-recipient's behaviour and personality changes. Patient apathy and communication difficulties were experienced as the most challenging behavioural issues for carers, as reported in qualitative studies (Johannessen et al., 2017; Massimo et al., 2013; Nichols et al., 2013; Oyeboode et al., 2013). Carers are often

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fearful of the care-recipients' risky behaviours and lack of ability to assess danger (e.g. such as in the case of driving), leading carers to worry about leaving them unattended (Johannessen et al., 2017; Massimo et al., 2013; Oyebode et al., 2013; Rasmussen et al., 2019). Behaviours take an emotional toll on carers, with qualitative and quantitative studies both reporting carers feeling guilty (Chow et al., 2011; Nichols et al., 2013; Rasmussen et al., 2019; Reidijk et al., 2009), distressed, exhausted (Johannessen et al., 2017; Nichols et al., 2013; Nicolaou et al., 2010; Massimo et al., 2013) and in some cases, depressed (Diehl-Schmid et al., 2013; Rasmussen et al., 2019; Riedijk, 2009; Roche et al., 2015).

Carer's experience depression at times, with scores from two quantitative studies (that used the same cohort) indicating that about 50 per cent of the carers showed symptoms of depression (Diehl-Schmid et al., 2013; Roche et al., 2015). Furthermore, depression was positively correlated with worsening behavioural symptoms and the carer and care-recipient relationship (Diehl-Schmid et al., 2013), and carer depression and financial difficulties were associated with intensity of carer needs (beta = .25; $p = .01$; Roche et al., 2015). Additionally, carers often have difficulty balancing carer duties alongside their own personal responsibilities (e.g. work and self-care), resulting in them needing to reduce their work schedule or take leave from work (Diehl-Schmid et al., 2013; Rasmussen et al., 2019, Riedijk et al., 2008, 2009), potentially contributing to further financial strain (Diehl-Schmid et al., 2013; Oyebode et al., 2013; Roche et al., 2015). One quantitative study reported that at a 24-month follow-up, carers who indicated that they often sacrificed their own personal life to care for the care-recipient showed greater psychological difficulties and reduced overall QoL (Riedijk et al., 2009).

Positive experiences and resilience. The majority of the studies focused on negative aspects of caring for a person with FTD, and only two quantitative studies and one qualitative study identified aspects of possible resilience or positive experiences associated with caring

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(Chow et al., 2011; Nichols et al., 2013; Riedijk et al., 2008). One qualitative study reported that children valued sharing in meaningful sentimental moments with the care-recipient (Nichols et al., 2013), a quantitative study reported that 20 per cent of carers discovered strength in dealing with the challenges (Chow et al., 2011), and findings of a longitudinal study reported that psychological well-being and QoL remained stable over a two-year period (Riedijk et al., 2008).

Coping. Coping strategies enabled carers to manage challenging aspects of caring (Massimo et al., 2013; Nichols et al., 2013; Oyeboode et al., 2013; Riedijk et al., 2008; Roche et al., 2015; Wong and Wallhagen, 2014). Coping strategies used amongst carers include separating the disease from the care-recipient, blaming the illness rather than the person (Massimo, Evans, & Benner, 2013; Nichols et al., 2013; Oyeboode et al., 2013) and acceptance that the person will never be the same (Massimo et al., 2013; Nichols et al., 2013; Oyeboode et al., 2013). Additional helpful coping strategies included: participating in hobbies (Oyeboode et al., 2013), educating others about FTD (Nichols et al., 2013; Oyeboode et al., 2013), spending quality time with care-recipients (Nichols et al., 2013; Oyeboode et al., 2013), using “white lies” or humour to intercept or defuse challenging behaviours (Oyeboode et al., 2013).

Quantitative studies evaluated the impact of coping strategies on carers. Two of these placed coping strategies into categories based on Lazarus and Folkman’s transactional theory of stress and coping (1984, 1991; Roche et al., 2015; Wong and Wallhagen, 2014). Carer experience suggests that coping may help to reduce the impact of carer strain and depression, as was reported in one study by Roche and colleagues (2015), who identified that coping mediated the relationship between carer strain and depression (coefficient 1/4 1.10, $p < .001$). Furthermore, emotional-focused coping (EFC) may improve couples’ satisfaction and carer mental health, as reported in the findings from Wong and Wallhagen (2014) study

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Support needs. Carers identify key support resources used, as well as areas needed for additional support (Chow et al., 2011; Diehl-Schmid et al., 2013; Johannessen et al., 2017; Nichols et al., 2013; Nicolaou et al., 2010; Rasmussen et al., 2019; Riedijk et al., 2008, 2009; Roche et al., 2015). These include the importance of raising public awareness about FTD and the needs of patients and families as well as educating health and social care professionals about FTD (Chow et al., 2011; Diehl-Schmid et al., 2013; Johannessen et al., 2017; Rasmussen et al., 2019). Carer's express the necessity for medical professionals to take initial concerns seriously at early stages of the disease, and for professionals to provide carers with psychosocial support (Rasmussen et al., 2019). Carer's additionally express the necessity of receiving support to enable them to care for the care-recipient at home initially, with opportunities for speciality day carers and a path to a specialty nursing home where appropriate (Johannessen et al., 2017; Nicolaou et al., 2010). Some carers expressed a need to be supported to develop coping strategies to help manage the progression of the disease (Riedijk et al., 2008; Roche et al., 2015), and for psychoeducation on self-care and methods to create time for their personal needs (Nicolaou et al., 2010; Riedijk et al., 2009), information on financial resources available (Diehl-Schmid et al., 2013), and age appropriate support groups for young carers (Nichols et al., 2013; Johannessen et al., 2017).

Helpful sources of support experienced by carers include: receiving information, emotional support, and sense of stability from family members (including carers' children) and friends, and healthcare professionals (Johannessen et al., 2017; Nichols et al., 2013; Oyebode et al., 2013). For some carers, receiving information is one of the most helpful means of support, followed by receiving psychosocial support by trained personnel, financial support, care assistance both inside and outside of the home and carer support groups, as reported in findings from one quantitative study (Diehl-Schid et al., 2013).

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Discussion

This is the first review to specifically examine the subjective experiences (positive and negative), specific needs, coping strategies and sources of support identified by informal carers of people with FTD. Synthesised findings from the review could facilitate the adaptation of currently available carer support services, and inform the development of specialised services that can meet the unique needs of carers of people with FTD. Six themes were reported to depict carers' experience and included: a) the challenging road to and receipt of diagnosis, b) relationship change and loss, c) challenging experiences in caring, d) positive experiences and resilience, e) coping, and f) support needs. Three of the most salient themes are discussed below.

The *challenging road to and receipt of diagnosis* highlights important aspects of the carer and patient experience and provides a greater understanding of the factors which influence the delay in diagnosis amongst people with FTD (Rosness et al., 2016). Findings from the review are in line with previous research, suggesting a delay in diagnosis, and the key role that carers have in reaching a diagnosis. This is due in part to the subtle and gradual onset of symptoms, which can make it difficult for carers to recognise symptoms and attribute them to symptoms of dementia (Ducharme et al., 2013; Rogers et al., 2017). Further this highlights the increased potential difficulty that individuals experiencing symptoms may have in accessing diagnosis when living alone or without a carer.

This theme extends the existing literature by highlighting the significant emotional impact of the pre-diagnostic stage on carers, suggesting that it is one of the most troublesome aspects of caring for a person with FTD and therefore a time when carers are likely to be in greatest need of information and public awareness regarding the symptoms of FTD (Chow et al., 2011; Diehl-Schmid et al., 2013; Johannessen et al., 2017; Rasmussen et al., 2019).

Therefore, there remains a great importance for healthcare professionals to be educated on the

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initial symptoms and presentations of FTD, so that informal carers and families may be able to address their concerns, and receive timely diagnosis and appropriate support (Rasmussen et al., 2019).

Findings from the review provide a deeper understanding of the FTD carers' experiences of *relationship change and loss*, which brings together literature that identifies a dramatic shift in relationships and identity (Bott et al., 2014; Ducharme et al., 2013; Mioshi et al., 2013; Pozzebon et al., 2016), and details a deep sense of loss experienced by carers who, as a consequence of FTD, often feels they have lost the person that they once knew (Diehl-Schmid et al., 2013; Massimo et al., 2013; Oyeboode et al., 2013; Rasmussen et al., 2019). This potentially highlights the need to address the construct of 'pre-death grief' (Lindauer & Harvath, 2014), whereby a person is grieving the loss of a person who is still living. Research suggests carers of people with dementia experience pre-death grief, and this can be associated with carer depression, burden and maladaptive coping (Lindauer & Harvath, 2014). Therefore it will be important for research to further develop an understanding of 'pre-death grief' to highlight important avenues for carer support.

Changes in the relationship and the need for people to take on increasing responsibilities as a carer can lead to a sense of isolation for a carer, further impacting on the carer and care recipient relationship and likely impacting both the carers' and care-recipients' quality of life (Diehl-Schmid et al., 2013; Johannessen et al., 2017; Massimo et al., 2013; Nichols et al., 2013; Oyeboode et al., 2013; Riedijk et al., 2008; Wong and Wallhagen, 2014). Further exploring how carers come to terms with loss and re-negotiate the change in relationship and identity may help to develop a deeper understanding of the relationship between the carer-patient dyad and carers' QoL (Diehl-Schmid et al., 2013; Riedijk et al., 2008). The review further emphasises the importance of receiving support to allow for carer

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respite (Nicolaou et al., 2010; Parahoo et al., 2002; Riedijk et al., 2009) as a way of improving the wellbeing of carer and care-recipient relationships.

Findings reported in the theme of *challenging experiences in caring* were in line with reports of carer burden and difficult experiences of carers reported in published reviews (Caceres et al., 2016; Karnatz et al., 2019; Baptiste et al., 2016). Findings from research suggests that carers experience difficulties adjusting to the profound personality and behavioural changes of bvFTD, which often impacts negatively on carer wellbeing. The review identified specific behavioural issues reported as most challenging and impactful on carers. These included apathy of the person with FTD, risky behaviours, lack of insight, and communication and interpersonal difficulties (Johannessen et al., 2017; Massimo et al., 2013; Oyeboode et al., 2013; Rasmussen et al., 2019). The current review highlighted that the impact of these difficulties often increases carer burden, while decreasing carer's psychological wellbeing. With increasing behavioural demands on carers, many are forced to leave work and may withdraw from their social environments (Diehl-Schmid et al., 2013; Rasmussen et al., 2019, Riedijk et al., 2008, 2009), leaving carers more vulnerable to experiencing isolation and financial strain. These factors are likely to exacerbate distress, exhaustion, and depression.

Therefore, findings of the current review highlight an increased need for carers of people with Frontotemporal dementia to receive education about FTD, training and resources to help them manage the symptoms of FTD, access to age appropriate support groups, coordination of care, and support to connect patients and families with services (Baptiste et al., 2013; Caceres et al., 2016; Cardarelli et al., 2010; Ducharme et al., 2013; Merrilees and Ketelle, 2010; Warren et al., 2013). Financial burden should further be addressed by providing resources to help carers meet the demands of their carer role (Cardarelli et al., 2010; Diehl-Schmid et al., 2013; Merrilees and Ketelle, 2010).

Critique of the literature

Firstly, key socio-demographic descriptors of the sample were missing from the included studies, which could provide important information regarding the ethnic and cultural factors likely to impact on the reported needs and helpful support services presented in the review. However, these limitations are likely to be influenced by the limited investigations into this topic area, and are common in other reviews of its kind (Caceres et al., 2016; Farina et al., 2017).

The identification of helpful coping strategies of carers of people with FTD was limited in this review, however two of the reviewed studies may highlight the important role of adaptive coping strategies in enabling carers to adapt to the caring role and the difficult changes in the carer-patient relationship, thereby improving carer outcomes of depression and QoL (Roche et al., 2015; Riedijk et al., 2008; Wong and Wallhagen, 2014). However, given the limitations of these studies, which highlight low internal consistency and possible response bias, results on coping should be interpreted with caution. Further research is needed to explore the function of coping strategies in the carer experience, and to explore possible areas of resilience in caring for people with FTD. Further exploration of positive, meaningful or valued experiences in caring could highlight important avenues for intervention and development of helpful coping strategies for carers of people with FTD.

Lastly, only one study explored the experience of children of people with FTD (Nichols et al., 2013), highlighting this as an important area for further research, develop and understanding of the unique experiences and needs of this population (Caceres et al., 2016). Additionally, no study in this review specifically addressed the experience of caring for children living in the home, a common experience of early onset dementias. Furthermore, although four studies reported the presence of child dependents in the home at the time of

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data-collection (Diehl-Schmidt et al., 2013; Johannessen et al., 2017; Nichols et al., 2013; Roche, et al., 2015), this does not include the impact of having children in the home during the early years of the disease, prior to diagnosis, given participants in some studies had been experiencing symptoms for more than 12 years. Furthermore, the genetic link of the disease was not addressed within the included articles, despite the significant challenges this holds for young families with children who have questions regarding family genetic testing and disclosure (Riedijk et al., 2009). Therefore future research should investigate the impact of caring for children or dependents, as a potentially significant aspect of the carer experience.

Strengths and Limitations

There are limitations to this review. No study was removed on the basis of quality, however results reported in the studies may be biased due to the use of snowball methodology, which used existing support networks to recruit for their studies, excluding those who did not seek support or those who experience barriers to accessing support. Therefore results of the review may be more representative of carers in greater need of support, which may explain the limited reporting of positive experiences of resilience in the carer experience, and may exclude the needs of people who did not access these supports.

A strength is the specificity of the review to the FTD experience through including only studies that do not combine FTD with other forms of dementia, or younger onset dementias to develop an understanding of the specific needs of this population.

Conclusions

Given the findings of this review, there is a pressing need for carers to be supported in developing coping strategies to help manage the unique behavioural symptoms of FTD (in bvFTD) as well as the unique experience of loss in the relationship caused by the disease. The lack of knowledge highlighted in this review means that it will be important to have

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public awareness campaigns, and to find ways to support carers in the pre-diagnostic stage to receive support from healthcare professionals through their journey.

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