**Cognitive rehabilitation, self-management, psychotherapeutic and caregiver support interventions in progressive neurodegenerative conditions: a scoping review**

Linda Clare1,2,3, Julia C. Teale1,2, Gill Toms4, Aleksandra Kudlicka1,2, Isobel Evans1, Sharon Abrahams5, Laura H. Goldstein6, John V. Hindle4, Aileen K. Ho7, Marjan Jahanshahi8, Dawn Langdon9, Robin Morris6, Julie S. Snowden10, Rhys Davies11, Ivana Markova12, Monica Busse13 and Jo Thompson-Coon2

1. Centre for Research in Ageing and Cognitive Health, University of Exeter
2. PenCLAHRC, University of Exeter Medical School
3. Centre for Research Excellence in Cognitive Health, Australian National University
4. School of Psychology, Bangor University
5. Psychology-PPLS, Euan MacDonald Centre for Motor Neurone Disease Research, The University of Edinburgh
6. King’s College London, Institute of Psychiatry, Psychology and Neuroscience
7. School of Psychology and Clinical Language Sciences, University of Reading
8. Institute of Neurology, University College London
9. Royal Holloway University of London
10. University of Manchester
11. Walton Centre NHS Foundation Trust
12. University of Hull
13. Cardiff University

Corresponding author

Professor Linda Clare, Centre for Research in Ageing and Cognitive Health (REACH), School of Psychology, University of Exeter, Perry Road, Exeter EX4 4QG, UK

l.clare@exeter.ac.uk

<http://www.psychology.exeter.ac.uk/REACH>

**Cognitive rehabilitation for people with progressive neurodegenerative conditions and cognitive disability: a scoping review**

**Abstract**

**BACKGROUND:** Despite their potentially significant impact, cognitive disability may be overlooked in a number of progressive neurodegenerative conditions, as other difficulties dominate the clinical picture.

**OBJECTIVE:** Weexamined the extent, nature and range of the research evidence relating to cognitive rehabilitation, self-management, psychotherapeutic and caregiver support interventions in Parkinsonian disorders, multiple sclerosis (MS), frontotemporal dementias (FTD), motor neuron disease and Huntington’s disease.

**METHODS:** Scoping review based on searches of MEDLINE and CINAHL up to 15 March 2016.

**RESULTS:** We included 140 eligible papers. Over half of the studies, and almost all the randomised controlled trials, related to MS, while a number of single case studies described interventions for people with FTD. CR interventions addressed functional ability, communication and interaction, behaviour or memory. The majority of psychotherapy interventions involved cognitive behavioural therapy for depression or anxiety. Self-management interventions were mainly available for people with MS. There were few reports of interventions specific to caregivers. Numerous methodological challenges were identified.

**CONCLUSIONS:** The limited range of studies for all conditions except MS suggests a need firstly to synthesise systematically the available evidence across conditions and secondly to develop well-designed studies to provide evidence about the effectiveness of CR and other psychological interventions.

**Keywords:** Parkinson’s disease, multiple sclerosis, frontotemporal dementia, motor neuron disease, Huntington’s disease, self-management, psychotherapy, caregiver

# Introduction

Progressive neurodegenerative conditions (PNCs), many age-related, affect over a million people in the UK alone and often result in cognitive disability. It is unrealistic to assume that there will be an early cure or means of prevention for all, or even most, progressive neurodegenerative conditions. Therefore, there is an urgent need for strategies that go beyond supportive care and enable people to function at the best level possible and to live well with cognitive disability resulting from neurodegenerative disease. Here we focus on five progressive neurodegenerative conditions in which cognitive disability, in some cases amounting to dementia, forms a significant part of the clinical picture. These are the Parkinsonian disorders (PDs) including Parkinson’s disease (PD), progressive supranuclear palsy (PSP), multiple system atrophy (MSA), corticobasal degeneration (CBD) and dementia with Lewy Bodies (DLB), multiple sclerosis (MS), the frontotemporal dementias (FTDs) including behavioural variant FTD, semantic dementia (SD) and primary progressive aphasia (PPA), motor neuron disease (MND), and Huntington’s disease (HD). We selected these five conditions, with a focus on the adult and older adult population, because they are amongst the commoner of the neurodegenerative diseases which apart from motor symptoms and behavioural changes are also characterised by significant albeit variable cognitive impairment. Alzheimer’s disease is not covered here because of the already extensively-reviewed research (Bahar-Fuchs, Clare, & Woods, 2013; Clare, 2008; Morris & Becker, 2004).

Cognitive impairment is integral to FTDs and HD, and affects a significant proportion of people diagnosed with MND, PDs, and MS. Cognitive impairments progress in extent and severity over time, although the rate of progression is variable across conditions and across individuals affected, and does not always correspond with the staging of physical symptoms. In FTDs, HD, and most PDs, cognitive impairments progress to full dementia (Hindle, Petrelli, Clare, & Kalbe, 2013), but this is rare in MS, and in MND occurs only where there is co-morbid FTD. Although the five conditions are very different in many respects, and have different pathogenic origins and affect different neural networks, there are common elements in terms of the underlying cognitive processes that are impaired. Table 1 summarises the cognitive domains affected and the behavioural and emotional features arising in each of the five conditions and demonstrates the extensive overlap in presentations.

(((Table 1)))

Impaired cognitive processes have wide-ranging effects. They may result in behavioural problems; for example, impaired executive function can involve poor judgement and decision-making, and can be associated with a loss of initiation and generation leading to apathy. Impaired cognition also contributes to the pathophysiology of motor symptoms; for example, problems with attention and conflict resolution contribute to mobility problems in PDs such as freezing of gait (Vandenbossche et al., 2011). Equally, motor symptoms may add to the cognitive challenges of daily life.

Cognitive changes, and the progression of cognitive impairment in some of these disorders, significantly affect important aspects of everyday life and well-being, including everyday functioning and behaviour (Birnboim & Miller, 2004; Goretti, Portaccio, Zipoli, Razzolini, & Amato, 2010; Goverover, Chiaravalloti, & DeLuca, 2016; Goverover, Haas, & DeLuca, 2016; Pirogovsky et al., 2014), employment (Benedict et al., 2016), communication and social interaction (Anderson, Simpson, Channon, Samuel, & Brown, 2013; Cavallo et al., 2011; Eddy, Mahalingappa, & Rickards, 2014; Meier, Charleston, & Tippett, 2010; Merrilees, Klapper, Murphy, Lomen-Hoerth, & Miller, 2010; Palmieri et al., 2010), involvement in decision-making (Abrahams, 2011; Martin et al., 2014; Meier et al., 2010; Merrilees et al., 2010), disease management (Bruce, Hancock, Arnett, & Lynch, 2010), and emotional well-being and quality of life (Ho, Gilbert, Mason, Goodman, & Barker, 2009; Ready, Mathews, Leserman, & Paulsen, 2008; Schrag, Jahanshahi, & Quinn, 2000; Shatil, Metzer, Horvitz, & Miller, 2010). Cognitive changes have a major impact on the family members of those living with these conditions, and cognitive impairment is associated with caregiver burden and reduced caregiver quality of life (Labiano-Fontcuberta, Mitchell, Moreno-García, & Benito-León, 2014; Morley et al., 2012; Ready et al., 2008), as are other psychiatric symptoms (Schrag, Hovris, Morley, Quinn, & Jahanshahi, 2006). Some people may lack awareness of changes or difficulties, especially in FTD and HD, which can add considerably to caregiver burden. Depression is common among people with HD, MS and PDs (Burn, 2002; Siegert & Abernethy, 2005; Slaughter, Martens, & Slaughter, 2001), and anxiety is also frequent in MS (Korostil & Feinstein, 2007), while people with FTDs and MND may also experience low mood (Blass & Rabins, 2009; Goldstein, Atkins, & Leigh, 2002).

Because of the predominance of other difficulties, such as progressive motor system dysfunction (PDs, HD, MS, and MND) or behaviour and personality change (FTD and HD), the impact of cognitive change in these conditions is often underestimated and under-recognised. Furthermore, with the exception of FTD, people living with these conditions who have cognitive impairments are often excluded from intervention studies (Goodwin & Lan, 2014). Consequently, there is limited guidance available that can inform clinicians and those living with or affected by these conditions about effective non-pharmacological ways of managing the impact of cognitive impairment and supporting optimal functioning. Appropriate pharmacological treatment where indicated is of course essential, but here we focus on non-pharmacological approaches.

Importantly, strategies for managing the effects of cognitive impairment and optimising functioning are not condition-specific. The application of common strategies can be tailored to the specific needs of the population and the individual, for example by making adaptations to account for motor or sensory limitations. This means that evidence from a range of conditions could potentially be assembled to provide professionals with practical guidance that enables them to work flexibly across conditions, drawing on the commonalities that can be observed.

Rehabilitation is key to managing cognitive and behavioural disability and optimising functioning. Rehabilitation for people with cognitive disability, termed ‘cognitive rehabilitation’ (CR) and sometimes also referred to as ‘reablement’, focuses on enabling each individual to function at his/her optimal level given the nature and extent of cognitive impairment, and to remain engaged with an appropriate social context (Beaumont & Morris, 2005). CR uses a goal-oriented approach to facilitate improved management of the consequences of functional disability, and has already been successfully applied to assist people with dementia due to Alzheimer’s or vascular disease (Bahar-Fuchs et al., 2013; Clare et al., 2013; Clare et al., 2010). Key elements are an individualised formulation, collaborative identification of specific realistic and meaningful goals, application of evidence-based behavioural strategies, and adjustment of goals and strategies in response to changes in the person’s condition. The rehabilitation therapist assesses the person’s current ability and the demands of the activity the person wishes or needs to do, identifies where mismatches arise, and collaboratively develops a therapy plan to support goal attainment. While rehabilitation goals may be very different depending on condition, stage and degree of severity, the overall aims of supporting independence and self-determination, and the repertoire of behavioural methods and techniques employed to support goal attainment, are likely to be similar.

Cognitive rehabilitation is complemented by three other key related psychosocial intervention approaches:

* Access to psychological therapies to address mental health needs such as anxiety or depression, and support the process of adjustment to living with a neurodegenerative condition. This may be a vital precursor to enabling participation in cognitive rehabilitation interventions.
* Providing information about the condition and encouraging an appropriate degree of self-management. This encompasses support for decision-making and planning for future care needs, including end-of-life care where relevant, and may include facilitating opportunities for social contact and engagement in enjoyable and meaningful activities. Greater understanding of a condition can facilitate engagement with rehabilitation.
* Support for caregivers. Effective support for caregivers includes provision of information, advice, emotional support and skills training, enabling them to support the process of cognitive rehabilitation and to provide optimal care that promotes the functioning, independence and self-determination of the person with cognitive impairment.

While pharmacological interventions play a part in the management of cognitive and behavioural difficulties, and can facilitate participation in non-pharmacological interventions, the indications for and efficacy of such treatments represent a complex and specialised topic that is beyond the scope of the present review. Here we focus on non-pharmacological approaches.

Drawing together the evidence for the efficacy of rehabilitation interventions, together with associated domains of self-management, psychotherapeutic and caregiver support interventions, across the five PNCs, could provide a basis for developing guidance for health and social care professionals, and information for patients and carers living with PNCs, and could identify gaps in knowledge to be addressed by further research. While reviews have examined the efficacy of specific interventions for one or more conditions (Croot, Nickels, Laurence, & Manning, 2009; Fernie, Kollmann, & Brown, 2015; Hindle et al., 2013; Langenbahn, Ashman, Cantor, & Trott, 2013; Rae-Grant et al., 2011) no review has yet integrated the evidence for cognitive rehabilitation and associated intervention approaches across all five conditions. Here we outline a scoping review (Arksey & O'Malley, 2005; Colquhoun et al., 2014; Levac, Colquhoun, & O'Brien, 2010; Pham et al., 2014) undertaken to determine the feasibility of this approach and its potential for improving the quality of support available to people with these conditions and their caregivers. We defined our review question as: ‘What is the extent, nature and range of the research evidence relating to the efficacy, experience and implementation of cognitive rehabilitation, self-management, psychotherapeutic and caregiver support interventions in PDs, MS, FTD, HD and MND?’

# Materials and methods

In line with recommended scoping review methods, we aimed to map the application of key intervention concepts, and types of evidence available, in relation to people living with the conditions of interest and their caregivers by systematically searching, selecting and synthesising existing knowledge (Arksey & O'Malley, 2005) and to consider the implications for practice, within the constraints imposed by budgets and time availability (Levac et al., 2010). We adopted a broad focus and considered a wide range of study designs in order to provide a descriptive overview of the available evidence (Pham et al., 2014). We consulted with people living with PNCs when formulating plans for the project.

We conducted searches in the MEDLINE and CINAHL databases on 15 and 23 January 2015, and updated searches on 15 March 2016, combining search terms designed to capture the relevant types of intervention and the conditions of interest. We selected these databases for pragmatic reasons, given time and budget constraints, as two of the largest and most widely-used sources likely to provide the most effective search results. Our target populations were people with one of the five PNCs or carers of people with one of the five PNCs. Target interventions were cognitive rehabilitation, psychological therapy or self-management, education or support interventions for people with one of the five PNCs, or education, skills training or support interventions for caregivers of people with one of the five PNCs. We considered any or no comparators, and any outcomes. Search terms covering populations and interventions are shown in table 2.

(((Table 2)))

Studies were included if a) participants had one of the five PNCs or were carers of people with one of these conditions; b) participants with PNCs who had cognitive impairment were included or not explicitly excluded; and c) the paper reported quantitative or qualitative data relating to a cognitive rehabilitation, psychological therapy or self-management, education or support intervention for people with one of the five PNCs, or an education, skills training or support intervention for caregivers of people with one of the five PNCs. For PDs and MS, searches were limited to the period from 2010 to the present due to the volume of records available for those conditions. Key studies known to the research team were also included.

Title, abstract and full-text screening was carried out by GT, JCT, AK and IE. The screening process was managed in EndNote ® Version 7. After merging search results from MEDLINE and CINAHL, duplicates were identified in Endnote first by using the default settings in ‘Find duplicates’ and then by adjusting the search criteria and inspecting reference lists ordered alphabetically by title and by author. At each stage of screening, each item was considered by at least two people working independently and any disagreements were discussed in order to reach a consensus. Where a lack of clarity remained, the item was referred to LC for a final decision. Eligible studies were grouped according to type as follows:

1. Cognitive rehabilitation interventions addressing cognitive or behavioural disability.
2. Psychological interventions addressing mood or adjustment to living with a neurodegenerative disorder.
3. Education, self-management and support interventions, including symptom management.
4. Education, skills training and support interventions for carers of people living with neurodegenerative disorder, such as carer support groups.

# Results

Figure 1 provides a flowchart of the process of study identification and selection. Our searches yielded 19,187 unique records, of which 2,508 were retained for abstract screening and 731 were selected for full-text assessment. At this stage 154 papers were not readily obtainable and of the remaining 577 records, 140 reported relevant studies. Over half of the included studies focused on MS (74 studies, 53% of total). FTD (27 studies) accounted for 19%, PDs (19 studies) for 14%, HD (10 studies) for 7% and MND (10 studies) for 7% of the total. The included studies for each PNC are summarised by research design and by topic in table 3.

(((Figure 1)))

(((Table 3)))

To prepare a descriptive overview, the identified studies were grouped into the four intervention categories: cognitive rehabilitation (63 studies), psychological therapies for adjustment and mood (28 studies), education and self-management (42 studies), and support for carers (7 studies). A descriptive summary of the included studies, grouped by category and condition, is provided in table 4. Following the review of each category, methodological issues and challenges are considered.

 (((Table 4)))

## Cognitive rehabilitation

The cognitive rehabilitation interventions described were conducted either with individuals, in groups, or as part of multi-component treatment programmes. Interventions focused on functional ability, communication and interaction, behaviour or memory. Examples of studies using this approach were found across all five conditions. Of the 21 studies found involving people with FTD and SD, most were single case designs. Sample sizes ranged from 1 – 9. In contrast the 21 studies in MS (Gentry, 2008; Lincoln, Dent, & Harding, 2003; Lincoln et al., 2002) included 8 randomised controlled trials and sample sizes ranged from 1 – 240. There were 7 studies in HD (Mattern & Kane, 2007), of which 5 evaluated the effects of inpatient multidisciplinary team interventions and 2 were single case designs. There were 9 studies of PDs (Jain, Dawson, Quinn, & Playford, 2004), including one pilot RCT and one multi-centre RCT. For MND, there were 4 case studies (Kaiser et al., 2001) and one between groups retrospective analysis including 344 people (Traynor, Alexander, Corr, Frost, & Hardiman, 2003).

*Individualised, goal-oriented interventions to support everyday functioning,* *activities of daily living and self-care* drawon compensatory methods such as the use of memory aids, restorative methods including techniques to help take in and recall information, such as spaced retrieval, and environmental adaptations (Buchanan, Christenson, Houlihan, & Ostrom, 2011). Studies examining the relative benefits of different techniques provide information about which strategies are most helpful for whom in which contexts (Goverover, Chiaravalloti, & DeLuca, 2008). Drawing on evidence about the efficacy of specific techniques and approaches, individual interventions are designed to address specific goals or needs, taking account of the individual’s profile of strengths and limitations in cognitive and functional ability. Examples of studies using this approach were found across all five conditions.

*Individualised, goal-oriented interventions to support communication* *and social interaction* include the introduction of specific information processing strategies as well as the use of assistive communication aids and compensatory strategies and development of non-verbal communication skills (Croot et al., 2009; Kortte & Rogalski, 2013). Use of communication aids to enhance interaction was described for people with HD (Klasner & Yorkston, 2001). Extensive research in the FTDs applied a range of techniques to support relearning or maintenance of semantic knowledge (Bier et al., 2009; Dewar, Patterson, Wilson, & Graham, 2009; Dressel et al., 2010; Jokel & Anderson, 2012; Jokel, Rochon, & Leonard, 2006; Newhart et al., 2009; Robinson, Druks, Hodges, & Garrard, 2009; Savage, Ballard, Piguet, & Hodges, 2013; Savage, Piguet, & Hodges, 2015; Snowden, Kindell, Thompson, Richardson, & Neary, 2012; Snowden & Neary, 2002; Suárez-González et al., 2015). Some studies considered maintenance of gains and transfer to everyday life (Bier et al., 2009; Robinson et al., 2009; Savage et al., 2013).

*Individualised, goal-oriented interventions for behaviour* are based on an individual formulation and on an understanding of the antecedents, consequences and functions of the given behaviour, and can help to reduce the impact of behavioural changes and hence support everyday functioning, relationships and social interactions. Techniques include introducing distraction and controlling triggers, as well as implementing environmental modifications, aids and adaptations (Croot et al., 2009). The use of this approach was reported for people with FTD and MND (Cleary, 2008; Fick, van der Borgh, Jansen, & Koopmans, 2014).

*Cognitive rehabilitation interventions* *for memory and other cognitive difficulties* were reported mainly in MS, often using a group format. These interventions, for people with mild to moderate impairments in memory, attention or executive function, encourage the use of compensatory and restorative strategies, environmental adaptations, and assistive technology. Included studies targeted self-efficacy and strategy use (Shevil & Finlayson, 2009, 2010; Stuifbergen et al., 2012) and subjective memory problems (Carr, das Nair, Schwartz, & Lincoln, 2014).

*Cognitive rehabilitation forms a component of some multi-disciplinary rehabilitation programmes.* Rehabilitation programmes conducted on an in-patient or out-patient basis may include a focus on addressing the consequences of cognitive impairments, although it is often not possible to determine the extent to which this contributes to the overall outcomes. Such programmes have been reported for example in HD, aiming to reduce disability and improve participation and quality of life (Frich, Røthing, & Berge, 2014; Piira et al., 2013; Thompson et al., 2013; Zinzi et al., 2007).

## Psychological interventions for adjustment and mood

The majority of studies were found in MS; of 17 included studies, 7 were RCTs. Fourteen studies evaluated the effects of cognitive behavioural therapy (Mohr et al., 2005) and 2 focused on acceptance and commitment therapy (Nordin & Rorsman, 2012). The majority of these were individual interventions, although there were also some group-based interventions addressing anxiety and depression, using approaches such as CBT, mindfulness, or acceptance and commitment therapy. There were 5 studies in PDs, including one RCT; all evaluated CBT interventions (Dobkin, Allen, & Menza, 2007; Dobkin et al., 2011; Dobkin et al., 2014) and all but one of these were delivered on a one-to-one basis. Three studies in MND included an RCT of individual psychotherapy (Averill, Kasarskis, & Segerstrom, 2013), a feasibility study of dignity therapy (Bentley, O'Connor, Kane, & Breen, 2014), and a case study of a CBT-based intervention (Pérez & Dapueto, 2014). Two case studies in HD reported a CBT (Silver, 2003) and a remotivation therapy (Sullivan, Bird, Alpay, & Cha, 2001) intervention respectively. Thus while CBT has been used to address mood and emotions in a range of PNCs (Fernie et al., 2015), other psychotherapeutic approaches, such as dignity therapy and remotivation therapy, have also been reported, although overall the evidence-base appears limited (Gould et al., 2015). There were no reports of psychotherapeutic interventions for people with FTD.

## Education and self-management

There were 36 studies in MS reporting education, self-management and support groups conducted either in person, via telephone or online, including 15 RCTs and a number of qualitative evaluations (Cecil, 2014; Regan et al., 2008). Four studies reported similar interventions for people with PD (Mulligan, Arps, Bancroft, Mountfort, & Polkinghorne, 2011) and one study surveyed 726 people about their experiences of using support groups (Dorsey et al., 2010). Two studies sought the views of people with MND attending a support group (Kasarkis, Elza, Bishop, & Spears, 1997; Regan et al., 2008). These surveys suggest that support groups may be accessed by significant numbers of people with some PNCs, but that outcomes are rarely evaluated. There were no reports of education, self-management or support groups for people with HD or FTD.

## Support for caregivers

Only a small number of studies examined the effects of interventions for caregivers of people with PNCs. These carer interventions covered a mixture of progressive neurodegenerative disorders: FTD, HD, PDs and atypical early-onset dementias (Mioshi, McKinnon, Savage, O’Connor, & Hodges, 2013). Five studies reported on the effects of participating in caregiver support groups. One study investigated whether participants were in contact outside of group meetings (Dipple & Evans, 1998). One study assessed the effects of a positive emotion skill-building intervention (Dowling et al., 2014).

*3.5 Methodological issues*

Relevant studies were identified across all five conditions and all four types of intervention. However, the distribution across conditions and types of intervention was highly skewed with almost half of all studies, and 89% of all RCTs, relating to MS. There was a lack of randomised controlled trials in relation to the rarer PNCs. In many cases participant characteristics were insufficiently detailed; descriptions of the type and degree of cognitive impairment were often limited or even lacking altogether. Interventions implemented in group studies and RCTs were often poorly-described, although in contrast the availability of numerous single-case designs meant that some interventions were clearly characterised. Interventions were also sometimes wrongly labelled with misleading names which did not properly reflect the nature of the intervention being reported. Reporting of case studies tended to focus on specific changes in performance relating to goals, needs or behaviours directly targeted in the intervention. In contrast, larger trials often reported only more distal measures such as quality of life or mood, covering areas where there may not necessarily be problems or impairments at baseline, or focused on measures of impairment such as neuropsychological tests which would not necessarily be expected to improve.

# Discussion

This scoping review shows that there is a body of evidence across the five PNCs, despite the relative rarity of conditions such as FTD, HD and MND (Bilney, Morris, & Perry, 2003; Langenbahn et al., 2013), the lack of emphasis on cognitive and behavioural disability relative to other aspects of the condition in PD, MS and MND (Hindle et al., 2013), and the frequent exclusion from trials of participants with any degree of cognitive impairment or psychiatric co-morbidity. The review yielded relevant studies across all five conditions and all four types of intervention. However, the distribution across conditions and types of intervention was highly skewed with a large proportion of studies relating to MS, where the depth and quality includes Class 1 evidence (Chiaravalloti, Moore, Nikelshpur, & DeLuca, 2013). The evidence for cognitive rehabilitation in MS was examined in a previously-published Cochrane review (Rosti‐Otajärvi & Hämäläinen, 2014). Thus, the need for further research is likely to be greater for the other four conditions.

The limited number of studies in PDs was somewhat surprising, given that PD itself is the most frequently-occurring PNC after Alzheimer’s disease, cognitive impairment is widespread, and a high proportion of people living with PD will eventually develop dementia. This may have resulted in part from the time constraints that were imposed on the search period due to the very large numbers of records identified, but is likely also to reflect the lesser emphasis placed on cognitive as opposed to motor symptoms in PDs. The nature of FTD, HD and MND make it difficult to gather large enough sample sizes for single-centre intervention trials, so it is to be expected that the majority of reports would focus on single-case designs. For these rare conditions, multi-centre studies will be required to demonstrate effectiveness of interventions at group level.

With regard to types of intervention, the largest group of studies reported cognitive rehabilitation interventions. There were relatively few studies of psychological interventions or of self-management, education or support. The progressive nature of the cognitive and behavioural changes in FTD and HD, as well as debilitating motor aspects in HD, may limit the perceived relevance of these approaches, while people with PDs who develop significant cognitive impairment are likely to have been living with the condition for some time. Additionally, however, many professionals and services providing psychological interventions will not have a specific remit to provide services to these groups, and may lack knowledge about these disorders and experience difficulty in adapting the interventions to take account of motor problems and other condition-specific constraints. There is a need to develop a clear understanding about which interventions are most appropriate for people with each condition and how these may best be adapted to ensure suitability and acceptability. There were surprisingly few studies of support for caregivers of people with these conditions; possibly some caregivers are accessing support from charities focused on the specific conditions while others may be accessing generic support for caregivers of people with dementia or a range of disabilities, but these approaches are not being evaluated systematically.

The findings highlight some of the key challenges inherent in attempting to synthesise evidence regarding psychosocial interventions, which must be addressed in order to provide definitive conclusions about efficacy and useful information for practitioners. These relate to study design, selection of participants, description of interventions, application of specific rehabilitation techniques, and choice of outcome measures. The lack of randomised controlled trials in relation to the rarer PNCs limits confidence in the robustness and generalisability of findings, especially as all the included studies reported some positive results, raising concerns about possible publication bias. Participants with PNCs are heterogeneous, and it is important to characterise the type and degree of cognitive impairment, and to take participant characteristics into account in order to provide information that is useful to practitioners. Similarly, clear descriptions of interventions are needed. It is important that interventions are correctly named and clearly described. Specific cognitive rehabilitation techniques and strategies could be applied in different ways across studies, making it especially important to derive appropriate guidance on how to incorporate these strategies into practice. In larger trials where evaluation focused on areas where participants might not have impairments or specific needs at baseline, or on measures of impairment which would not necessarily be expected to improve, the trial design could yield a negative outcome for a potentially-effective intervention. It is important to acknowledge that in a progressive condition an outcome of no change or slowed deterioration can indicate treatment effectiveness (Croot et al., 2009).

This review has a number of limitations. In undertaking the review, pragmatic considerations regarding budget and time available had to be balanced against achieving sufficient coverage and breadth (Levac et al., 2010). More comprehensive searches and the ability to access papers that were not readily available would yield additional studies for inclusion; however, while we may not have accessed the full extent of the evidence, we believe that the scoping searches have provided a reasonably comprehensive picture of the nature and range of the evidence available. We have not undertaken any formal assessment of the quality of individual studies, but this is in line with the procedures adopted in other scoping reviews (Pham et al., 2014), where the aim is to gain an overview of the evidence rather than to make a detailed assessment of individual studies. We considered that given the preliminary nature of this review, an evaluation of study quality would not have added significantly to the work.

# Conclusions

This scoping review has described the extent, nature and range of the research evidence relating to the effectiveness, experience and implementation of cognitive rehabilitation, self-management, psychotherapeutic and caregiver support interventions in PDs, MS, FTD, HD and MND. The limited range of studies for all conditions except MS suggests it will be important firstly to systematically synthesise the available evidence across these conditions, taking account of the methodological challenges inherent in this endeavour, and secondly to build on this by developing rigorous, well-designed studies to provide robust evidence about the effectiveness of CR and other psychological interventions in PDs, FTD, HD and MND. There is a need for evidence that can provide a basis for developing guidance to support the practice of health and social care professionals, and advice for people affected by these conditions and their carers, that could enhance the possibility of living well with cognitive disability.

**Declaration of interest statement**

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**Table 1. Cognitive, behavioural and emotional features that may arise in each of the five PNCs**

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Possible features:** | PDs | MS | FTDs | HD | MND |
| Executive function impairment | √ | √  | √ | √ | √ |
| Attention impairment | √ |  | √ | √ | √ |
| Information processing impairment | √ | √ | √ | √ |  |
| Working memory impairment | √ |  | √ | √ | √ |
| Episodic memory impairment | √ | √ | √ | √ | √ |
| Language and semantic impairment |  |  | √ | √ | √ |
| Visuospatial perception impairment | √ |  |  | √ |  |
| Delusions and hallucinations | √ |  | √ | √ | √ |
| Behavioural issues | √ |  | √ | √ | √ |
| Apathy | √ | √ | √ | √ | √ |
| Depression and anxiety | √ | √ | √ | √ | √ |

**Table 2. Search terms used in the scoping review**

Population-related search terms

Huntington\*

“Frontotemporal Dementia\*” OR “Behavioural Variant Frontotemporal Dementia\*” OR “Semantic Dementia\*” OR “Progressive Non Fluent Aphasia\*” OR “Progressive Non-Fluent Aphasia\*” OR “Frontotemporal Logopenic Aphasia\*” OR “bevFTD” OR “FTD”

“Motor Neuron” OR MND OR ALS OR “Amyotrophic Lateral Sclerosis” OR “Lou Gehrig\*” OR “Progressive Bulbar Pals\*” OR “Progressive Muscular Atroph\*” OR “Primary Lateral Sclerosis” OR “Anterior Horn Cell\*” OR “Mill\* Syndrome\*”

“Multiple Sclerosis” OR “Clinically Isolated Syndrome” OR “Radiologically Isolated Syndrome”

Parkinson\* OR “Progressive Supranuclear Palsy” OR “Multiple System Atrophy” OR “Corticobasal Degeneration” OR “Lewy Body Dementia\*”

“Progressive Supranuclear Palsy” OR “Multiple System Atrophy” OR “Corticobasal Degeneration” OR “Lewy Body Dementia\*”

Intervention-related search terms

Intervention\* OR Treatment\* OR Therap\* OR “Randomised Control\* Trial\*” OR “Intervention\* Stud\*” OR Rehabilitation OR Remediation OR Retraining OR “Single case\*” OR “Support group\*” OR Psychoeducation OR Education OR “Self-management” OR Advice

**Table 3. Overview of studies included in the scoping review for each PND**

1. **By research design**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
|  | MS (n=74) | PDs(n=19) | FTD (n=27) | HD (n=10) | MND (n=10) | Total (n=140) |
| RCT  | 31 | 3 | 1 | 0 | 0 | 35 |
| Secondary analysis of RCT  | 11 | 1 | 0 | 0 | 2 | 14 |
| Non-randomised group designs | 22 | 10 | 3 | 5 | 2 | 42 |
| Single case designs or case series | 3 | 1 | 21 | 3 | 4 | 32 |
| Qualitative evaluation | 7 | 4 | 2 | 2 | 2 | 17 |

1. **By type**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
|   | MS (n=74) | PDs(n=19) | FTD (n=27) | HD (n=10) | MND (n=10) | Total (n=140) |
| Cognitive rehabilitation | 21 | 9 | 21 | 7 | 5 | 63 |
| Psychotherapy | 17 | 5 | 1 | 2 | 3 | 28 |
| Self-management | 36 | 4 | 0 | 0 | 2 | 42 |
| Carer interventions | 0 | 1 | 5 | 1 | 0 | 7 |

**Table 4. Studies included in the review**

1. **Cognitive rehabilitation**

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Authors & Date** | **Design** | **Sample** | **Intervention** | **Aims** | **Key findings** |
| **Fronto-temporal dementia (FTD) including semantic dementia (SD) – 22 studies** |  |  |  |  |  |
| Bier et al. (2009) | Case study, pre-post- follow up | 1 person with SD, 5 controls  | Spaced retrieval  | Explore formal semantic therapy | Better on trained items. Maintained at 5 weeks. No generalisation.  |
| Bier et al. (2011) | Case study, pre- post- follow up | 1 person with SD | Ecological approach CR | Explore effectiveness | Cooked more independently and initiating more cooking.  |
| Bier, Brambati, et al. (2015)  | Case study | 1 person with SD | Ecological CR  | Assess ability to learn new technologies | At 6-month follow-many technology functions still used. |
| Bier, Paquette, and Macoir (2015)  | Case study | 1 person with SD | CR  | Assess baseline use of compensations and ability to learn new technologies | With an errorless learning approach, he learnt to effectively use 10 smartphone functions. |
| Dewar et al. (2009) | 3 cases with HSE, 1 case with SD, pre-post | 4 people with SD | Mnemonics and errorless learning for relearning semantic information | Explored relearning of semantic knowledge | SD improved on trained items. Maintained at 1 month. some generalisation |
| Dressel et al. (2010) | 1 case, pre-post-follow up  | 1 person with SD | Phonological and semantic cueing hierarchies  | Investigate use of treatment | Improved with both techniques at 2 months but gains lost over time |
| Fick et al. (2014)  | Case report | 1 person with FTD | Behavioural intervention | Describe intervention for pacing + vocally disruptive behaviour | Vocally disruptive behaviour + pacing reduced |
| Frattali (2004)  | Case study, pre- post- follow up | 1 person with FTD | Errorless learning  | Explore effectiveness | Improved on trained, no generalisation, gains lost at 3 months |
| Izzo, DiLorenzo, and Roth (1986) | Case study | 1 person with Progressive Supranuclear Palsy | Therapeutic rehabilitation techniques. | Discuss the rehabilitation problems and management of the person. | Improvement in dysphagia and visual scanning. |
| Jefferies, Bott, Ehsan, and Ralph (2011) | Case study, between groups | 1 person with SD, 8 controls  | List recall and errorless learning  | Explore whether and how can learn phonological information | Phonological learning improved  |
| Jokel and Anderson (2012)  | 7 cases, pre-post | 7 people with SD | Errorless, errorful, active and passive learning of naming and comprehension of treated words  | Examine different combinations of training techniques | All training resulted in improvement. Errorless training was better maintained |
| Jokel et al. (2006)  | Case study, pre-post- follow up.  | 1 person with SD | Home treatment programme (practising words of objects) | Explore a treatment programme. Designed jointly with participant.  | Better on words with some retained semantic knowledge. Practice delayed progression of loss.  |
| Jokel, Rochon, and Anderson (2010) | 1 case, pre-post-follow up | 1 person with SD | Computer delivered errorless learning  | Explore effectiveness + feasibility | Good learning and 3 month retention |
| Newhart et al. (2009)  | 2 cases, pre-post (1 LPA, 1 SD) | 1 person with SD, 1 person with logopaenicprogressive aphasia | Cue hierarchy treatment for naming objects  | Compare treatment across LPA and SD on lexical processing assessment  | SD better with trained words but limited maintenance |
| O'Connor et al. (2016)  | Case study | 1 person with FTD-behavioural, 1 person with FTD-semantic | OT  | Describe intervention and assess feasibility | Engagement in more activities, improvements in behaviour, reduced caregiver distress |
| Robinson et al. (2009)  | Compared 2 cases with matched controls, pre- post- follow up | 2 people with SD, 2 controls | Errorless learning and training | Explore training effectiveness (object naming, definition and use) | Improved naming, definitions and object use |
| Savage et al. (2013)  | 6 cases, pre-post | 4 people with SD | Word retraining (repetitive practice of word-picture pairing) | Evaluate the success of training | More practice time improved maintenance (not much maintenance).  |
| Savage, Piguet, and Hodges (2014)  | Pre- and post-intervention comparison | 5 people with SD | Online word training program | Examine whether gains in training generalise from picture naming to everyday settings | All 5 improved naming the trained pictures; 4 out of 5 improved on video description task |
| Savage et al. (2015) | Case series of single-subject experimental design | 9 people with SD | Computer delivered rehearsal (individually tailored) of 75-100 words | Examine independent and assisted maintenance of object words  | 80% trained words retained at six months with independent or continuous revision |
| Snowden and Neary (2002) | Two case studies | 2 people with SD | Repetition (some elements of errorless learning) of object names  | Examine the impact of residual word/ object knowledge and contextual information on relearning | Partial semantic knowledge and experiential contextual information aided relearning |
| Snowden et al. (2012)  | Case study, pre-post- follow up | 1 person with PPA | Speech and language therapy | Case description | Improvement at 12 months but gains lost at 2 years |
| Suárez-González et al. (2015) | Case study. Compared standard and conceptual enrichment training | 1 person with SD  | Conceptual enrichment training | See whether generalisation improved if personalised semantic memories recruited | Better naming and generalisation with conceptual enrichment training |
| **Huntington’s disease (HD) – 7 studies** |  |  |  |  |  |
| Ciancarelli, Tozzi, and Carolei (2013)  | Repeated measures | 34 people with HD | Inpatient multifunctional neuro rehabilitation | Explore intervention effectiveness | Improvements lost at 3 month follow-up |
| Frich et al. (2014)  | Qualitative interview (patients and caregivers) or focus groups (professionals) post-intervention | 9 carers, 11 people with HD, 15 healthcare professionals | Group-based multidisciplinary residential rehabilitation (inc. training of cognitive function, assistive devices; individual counselling) | Assess feasibility (opinions of patients, caregivers, and professionals) | Patients and caregivers emphasised positive mental, physical, and social outcomes, professionals indicated intervention was feasible |
| Klasner and Yorkston (2001)  | Case study | 1 person with HD | SALT (linguistic and cognitive strategies taught to aid communicative participation) | Describe the use of linguistic and cognitive supplementation strategies | Improved frequency and satisfaction with communication and ADL completion |
| Mattern and Kane (2007)  | Case study | 1 person with HD | Occupational therapy- activity programme  | Describe the case | Improved sense of purpose and developed friendships |
| Piira et al. (2013)  | Repeated measures (3 admissions of 3 weeks over 1 year) | 37 people with HD | Inpatient rehabilitation (physical exercise, social activities, group sessions) | Replicate Zinzi et al 2007 study | Improved gait, balance, physical QOL, anxiety and depression. No deterioration in cognition |
| Thompson et al. (2013)  | Between groups | 20 people with HD | MDT OT rehabilitation  | Pilot the programme | Improvement in mobility, body strength, written errors and fat composition. Minor improvement cog + depression |
| Zinzi et al. (2007)  | Repeated measures (3 week intensive, 3 times a year) | 40 people with HD | Multicomponent- inpatient rehabilitation  | Explore effectiveness | Improved motor performance, ADL performance and depression |
| **Motor Neuron disease (MND) – 5 studies** |  |  |  |  |  |
| Cleary (2008)  | Case study | 1 person with MND | Behavioural – to decrease risk of pneumonia (e.g. instruction in good oral hygiene)  | Describe intervention | Improved nutrition, energy, satisfaction, reduced distress and improved caregiver QOL |
| Kaiser et al. (2001)  | Case study | 2 people with MND | OT- computer device training | Evaluate the training technique (locked-in patients learned to use an EEG-based communication device) | Learnt to control cortical potentials without continuous feedback |
| Kübler et al. (2001)  | Case study | 2 people with MND | OT- computer device training | Describe and evaluate training (locked-in patients learned to use an EEG-based communication device) | Able to learn to control amplitude of slow cortical potentials |
| Neumann, Kübler, Kaiser, Hinterberger, and Birbaumer (2003)  | Case study | 1 person with MND | OT- computer device training (brain-computer communication) | To describe how a participant controlled EEG potentials | Used mental strategies to shift EEG potentials |
| Traynor et al. (2003) | Between groups- retrospective analysis | 344 people with MND | MDT team care (neurologists, specialist nurses, occupational, speech therapists, pulmonologist, nutritionist, psychologist, social worker) | Evaluate outcomes on survival | MDT care increased survival time for bulbar onset patients (compared with general neurology clinic control) |
| **Parkinsonian Disorders (PDs) – 8 studies** |  |  |  |  |  |
| Clarke et al. (2016)  | Multi-centre RCT (PD REHAB Trial) | 762 people with PD | People with limitations in ADL were randomised to physiotherapy and occupational therapy or no therapy | Evaluate efficacy | There were no significant improvements in the activities of daily living measure or the Parkinson Disease self-report measure in the therapy group.  |
| Disbrow et al. (2012)  | Between groups, 10-day training  | 30 people with PD, 21 controls | Computerised cognitive rehabilitation (designed to improve motor-related exec function) | Evaluate efficacy | Improvement in motor initiation and some generalisation to cognitive switching. Improvement on Trail Making B-A |
| Gil-Ruiz et al. (2013) | Case study | 1 person with DLB | Environmental adaptation for management of the ‘mirror sign’ (inability to recognise one’s reflection)  | Case description | Improved DSM symptoms |
| Giovannetti, Seligman, Britnell, Brennan, and Libon (2015)  | Between-groups | 20 people with PDD, 20 people with AD, 20 people with PD | Goal cues (remind task goals/ improve performance on everyday tasks) | Assess feasibility | AD patients showed a significantly greater response to goal cues as compared with PD + dementia, the goal cues facilitated task completion, but did not promote error correction |
| Jain et al. (2004)  | Pilot RCT | 17 people with MSA  | Individual OT | Identify feasibility and preliminary impact | Improved ADL functioning and health related QOL |
| Meek et al. (2010) | Article describes the process of designing and recording the intervention | 19 people with PD | Individual OT to optimise independence  | Detail the intervention | Mean number of visits 5.7Found to be feasible and acceptable, informed larger phase 3 trial (PD REHAB) |
| Pretzer-Aboff, Galik, and Resnick (2011)  | Repeated measures | 21 people with PD | Individual function focused care intervention  | Test feasibility and impact | Improved time spent exercising, disability, impairment, outcome expectations for exercise and QOL |
| Sturkenboom et al. (2013) | Process analysis | 43 people with PD, 43 carers, 7 healthcare practitioners | Individual OT | Evaluate feasibility | Feasibility demonstrated but response rate 23% |
| **Multiple sclerosis (MS) – 21 studies** |  |  |  |  |  |
| Allen, Longmore, and Goldstein (1995) | Repeated measures- case study | 1 person with MS | Cognitive training (teaching mnemonic strategies to increase list learning + recall of names of faces) | Describe effect | Improved list learning, quicker name learning  |
| Allen, Goldstein, Heyman, and Rondinelli (1998) | Single group pre- and post-training comparisons | 10 people with MS | Computer-assisted teaching of imagery-based mnemonic strategies | Evaluate feasibility | Participants quickly learned the strategies but methods showed no significant effects on recall |
| Asano, Preissner, Duffy, Meixell, and Finlayson (2015)  | Participants taken from an RCT (Finlayson et al., 2005) appears to report only the goal-setting group | 81 people with MS | Goal-setting | Describe goals set and evaluate the extent to which participants keep goals over time | Around 50% of goals are achieved, the most common being IADL’s and short-term goals |
| Birnboim and Miller (2004) | Single group pre- and post-training comparisons and 2 case studies | 10 people with MS | Cognitive rehabilitation with metacognitive therapeutic approach for executive function deficits | Explore efficacy | An overall improvement |
| Carr et al. (2014)  | Pilot RCT | 48 people with MS | Cognitive rehabilitation  | Assess feasibility | Improved mood. No effect of memory programme on memory outcome measures  |
| Chiaravalloti et al. (2013) | RCT (double-blind, placebo) | 86 people with MS | Cognitive rehabilitation (modified Story Memory Technique, mSMT) | Treat learning impairment | Significantly improved learning and memory compared to placebo group |
| Chiaravalloti, Wylie, Leavitt, and Deluca (2012)  | RCT (double-blind, placebo) | 16 people with MS | Cognitive rehabilitation (modified Story Memory Technique, mSMT) | Investigate if the cognitive intervention created neural changes | Increased activation and improved list learning  |
| Ernst et al. (2013) | Repeated measures | 25 people with MS, 35 controls  | Cognitive training (autobiographical memory) | Probe the programme | Improved autobiographical memory performance |
| Eyssen et al. (2013) | Cluster RCT | 269 people with MS | Client-centred occupational therapy | Assess efficacy | No difference in primary outcome, negative findings in secondary outcomes, and positive findings in process outcomes |
| Fink et al. (2010)  | Between groups | 40 people with MS-RR | Cognitive training – attention/ exec function (RehaCom software) | Evaluate efficacy | Improvement in aspects of executive function, improved verbal learning |
| Gentry (2008)  | Repeated measures | 21 people with MS | Assistive Device training (personal digital assistants, e.g. calendar, alarms) | Evaluate impact on functional performance | Improved functional performance and satisfaction |
| Goverover et al. (2008)  | Between groups | 20 people with MS, 18 controls | Cognitive rehabilitation (self-generation to improve learning/ memory of meal preparation and managing finances).  | Examine strategy utility | Self-generation improved learning and memory recall  |
| Klaren, Hubbard, and Motl (2014)  | Pilot RCT, pre- and post- trial questionnaire | 70 people with MS | Behavioural intervention (to reduce sedentary behaviour) | Examine efficacy | Sitting time significantly reduced in the intervention group |
| Lexell, Flansbjer, and Lexell (2014) | Retrospective study with a pre-post design | 43 people with MS | Individualised, goal-oriented interdisciplinaryrehabilitation | Assess self-perceived performance and satisfaction with performance of dailyactivities | Self-perceived performance and satisfaction higher after rehabilitation program |
| Lincoln et al. (2002)  | RCT (single blind) | 240 people with MS | Cognitive rehabilitation (cognitive deficits identified with battery, then given tailored intervention. E.g. training in use of diaries) | Evaluate benefit cognitive assessment and intervention | No significant outcomes on mood, quality of life, subjective cognitive impairment, independence |
| Lincoln et al. (2003)  | Secondary analysis of RCT treatment group (Lincoln et al., 2002) | 29 people with MS | Cognitive rehabilitation (individualised) | Evaluate rehabilitation | Only a quarter of participants benefitted |
| Mäntynen et al. (2014) | RCT | 102 people with MS-RR and subjective and objective attentional deficits | Cognitive rehabilitation (inc. computer WM training, psychoeducation, strategy learning, psych support) | Evaluate efficacy | No improvement in cognitive performance (processing speed, SDMT). Patients perceived reduction in cognitive deficits.  |
| Rosti-Otajärvi, Mäntynen, Koivisto, Huhtala, and Hämäläinen (2013) | Secondary analysis of RCT (Mäntynen et al., 2014)  | 98 people with MS-RR | Neuropsychological rehabilitation (attention retraining, teaching compensatory strategies, psych support, homework).  | Evaluate factors related to outcome. | More benefit for those with more severe attentional deficits (+male) |
| Salminen, Kanelisto, and Karhula (2014) | Qualitative analysis of focus group data | 68 people with MS | Multi-professional group-based out-patient rehabilitation programs | Identify which components of rehabilitation are seen as helpful by people with MS | Diverse range of components identified  |
| Shatil et al. (2010)  | Between groups (unprompted, home-based, computerised training) | 107 people with MS | Cognitive training (CogniFit Personal Coach) | Examine impact on cognitive performance | Improved general memory, visual memory, verbal-auditory working memory, LTM retrieval, naming, recall and processing speed |
| Stuifbergen et al. (2012)  | RCT (single blind, waiting list control) | 61 people with MS | Cognitive rehabilitation (Memory, Attention and Problem Solving Skills for Persons with MS, MAPSS).  | Refine and test the intervention | Improved self-efficacy and frequency of compensatory strategy use |

**(b) Psychological therapies for adjustment and mood**

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| **Authors & Date** | **Design** | **Sample**  | **Intervention** | **Aims** | **Key findings** |
| **Fronto-temporal dementia (FTD) – 1 study** |  |  |  |  |  |
| Yokota et al. (2006) | Pre- and post-intervention comparisons | 8 people with FTD | Group-Home Care  | Evaluate impact of homelike physical and social environment in a care home. | Beneficial effects on behavioral and psychological symptoms of dementia, and psychotropic drug |
| **Huntington’s disease (HD) – 2 studies** |
| Silver (2003)  | Repeated measures, case study | 1 person with HD | CBT- individual | Describe key features of therapy and outcome | Improved anxiety and depression ratings |
| Sullivan et al. (2001)  | Case studies | 6 people with HD | Remotivation (group) therapy | Present case studies | Positive gains reported |
| **Motor neuron disease (MND) – 3 studies** |
| Averill et al. (2013)  | RCT | 48 people with MND | Psychotherapy | Examine impact on psychological well-being | Improved psychological well-being but not maintained at six months |
| Bentley et al. (2014)  | Repeated measures | 29 people with MND | Psychotherapy (dignity therapy) | Assess feasibility, acceptance and potential effectiveness | No significant outcomes |
| Pérez and Dapueto (2014)  | Case study | 1 person with MND | Computer assisted (augmentative-alternative communication technology) Psychotherapy (based on CBT) | Describe how therapy was provided | Positive outcomes reported (improved communication with computer device, improved self-esteem, lower distress) |
| **Parkinsonian Disorders (PDs) – 5 studies** |
| Dobkin et al. (2007) | Pilot repeated measures | 15 people with PD | Individual CBT for depression in PD | Feasibility testing |  |
| Dobkin et al. (2011) | RCT | 80 people with PD | Individual CBT for depression in PD | First trial of CBT in PD |  |
| Dobkin et al. (2014)  | Additional analysis of RCT | 80 people with PD | CBT | Describe neuropsychological outcomes |  |
| Fitzpatrick, Simpson, and Smith (2010) | Qualitative (semi-structured interview) | 12 people with PD | Mindfulness based cognitive therapy (MCBT) | Explore experiences of MCBT (8 week course) | Positive experiences/ gains reported |
| Jiménez-Murcia et al. (2012) | Between groups retrospective analysis (pilot) | 60 people with PD | Individual CBT for pathological gambling in PD | Characterise therapy response | No differences in response between pathological gamblers with or without PD |
| **Multiple sclerosis (MS) – 17 studies** |  |  |  |  |  |
| Askey-Jones, David, Silber, Shaw, and Chalder (2013) | Repeated measures (not blinded) | 49 people with MS | CBT (individual) for common mental disorders in MS  | Examine effectiveness | Reduced depression and anxiety but not maintained |
| Beckner, Howard, Vella, and Mohr (2010)  | Secondary analysis of RCT (Mohr et al. 2005) | 127 people with MS | CBT (telephone) for depression in MS | Examine whether social support is a mediator | Patients with higher social support responded better to CBT |
| Boeschoten et al. (2012) | Repeated measures (pilot) | 44 people with MS | CBT (computerised) for depressive systems in MS | Examine feasibility and to examine outcome | Improved depression, problem solving skills and anxiety |
| Bombardier et al. (2013)  | RCT | 92 people with MS | Telephone-based physical activity counselling for major depression in MS  | Examine effectiveness | Improved depression |
| Burns, Siddique, Fokuo, and Mohr (2010) | Secondary analysis of RCT (Mohr et al. 2005) | 102 people with MS | CBT (telephone) for depression | Effects of comorbid anxiety on outcome | Comorbid fear reduced maintenance and comorbid GAD increased anxiety symptoms |
| Clancy, Drerup, and Sullivan (2015)  | Single group pre- and post- intervention evaluation | 11 people with MS | CBT | Evaluate efficacy | Improvements in insomnia, fatigue, and depression, although symptoms persisted |
| Dennison, Moss-Morris, Yardley, Kirby, and Chalder (2013)  | Qualitative interviews | 30 people with MS | CBT (mixed format) for adjustment to MS | Gain insight into participant experience | Positive outcomes reported |
| Graziano, Calandri, Borghi, and Bonino (2014) | RCT | 82 people with MS | CBT (group) for reducing depression and improving quality of life  | Evaluate effectiveness | Improved QOL and (in men) psychological well being |
| Mohr, Classen, and Barrera (2004)  | Clinical trial, between groups | 63 people with MS-RR | CBT, supportive expressive group psychotherapy, or sertraline | Examined the effects of treatment for depression on social support | All treatment interventions alleviated depression and increased perceived social support, utilised social support, and satisfaction with support.  |
| Mohr, Hart, and Goldberg (2003)  | Pre- and post- intervention assessment | 60 people with MS-RR | Individual CBT, group psychotherapy, or sertraline | Examine the effects of treatment for depression on fatigue | All treatment interventions reduced fatigue severity, which was associated with fewer symptoms of depression.  |
| Mohr et al. (2005) | RCT + 12 month follow-up | 127 people with MS | CBT (telephone) and supportive emotion-focused therapy (telephone) for depression in MS | Evaluate efficacy for attention and nonspecific therapy effects | Significant improvement in depression measures and positive affect |
| Mohr, Hart, and Vella (2007) | Additional analysis of RCT (Mohr et al., 2005) | 127 people with MS | CBT (telephone) and supportive emotion-focused therapy (telephone) for depression in MS | Evaluate effects on disability and fatigue | CBT superior in reducing disability and fatigue, depression |
| Nordin and Rorsman (2012)  | Pilot RCT.  | 21 people with MS | Acceptance and commitment therapy (group)/ relaxation therapy (group) for MS with depression/ anxiety | Evaluate effectiveness | At 3 month follow up, relaxation training group had improvement in anxiety, acceptance and commitment therapy group showed improvement in self-rated acceptance |
| Rigby, Thornton, and Young (2008)  | RCT assessing outcomes at five points over a one year follow up | 138 people with MS | CBT (group), educational material (booklet), or educational material (booklet) + non-structured social discussion | Document mood, self-efficacy, and resiliency following group CBT and to examine whether benefits were greater than those derived from provision of education or group social interaction. | CBT improved self-efficacy and resiliency, and reduced anxiety, in comparison with educational booklet intervention (but no significant difference). Also no difference between educational book and educational book + social discussion |
| Sheppard, Forsyth, Hickling, and Bianchi (2010)  | Single group, pre- and post- assessment + 3 follow up | 15 people with MS | Acceptance and commitment therapy | Investigate efficacy | Significant improvement in depression, thought suppression, impact of pain on behaviour, and QOL, but no change in the experience of physical symptoms and mindfulness practice. |
| Thomas et al. (2013) | Multicentre RCT (not blinded) | 146 people with MS | CBT (group) for fatigue management | Effectiveness | Improved fatigue, severity and self-efficacy |
| van Kessel et al. (2008) | RCT | 72 people with MS | CBT or relaxation training | Assess efficacy | CBT group reported significant reductions in fatigue, but there was no difference in stress, mood, or fatigue related impairment. |

1. **Education and self-management**

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| **Authors & Date** | **Design** | **Sample** | **Intervention** | **Aims** | **Key findings** |
| **Motor neuron disease (MND) – 2 studies** |  |  |  |  |  |
| Kasarkis et al. (1997) | Survey | Approximately 100–130 people with MND attended at least one session | Informational support group using interactive video format | Describe the approach | High degree of user satisfaction and proficiency in using interactive video |
| Regan et al. (2008)  | Survey | 15 people with MND | Education and support group | Six month pilot of the programme | Positive feedback |
| **Parkinsonian Disorders (PDs) – 4 studies** |  |  |  |  |  |
| A’Campo, Spliethoff-Kamminga, Macht, Roos, and Consortium (2010)  | Repeated measures & feedback | 64 people with PD, 46 carers  | PEPP patient education | Evaluate caregiver impact | Improved burden (PD & caregiver) |
| A’Campo, Spliethoff‐Kamminga, and Roos (2011)  | Secondary analysis of A’Campo et al. (2010), standardised patient education programme for patients with Parkinson’s disease and their carers | 64 people with PD, 46 carers | PEPP group patient education  | Identify treatment effect modifiers | No modifiers of PD outcomes Potential predictor of treatment benefit – caregiver having higher cog function |
| Dorsey et al. (2010) | Postal survey | 726 people with PD | Support groups (patient) | Assess patient satisfaction with medical care and use of support groups | 61% reported attending support group, 49% satisfied |
| Mulligan et al. (2011)  | Single group post-intervention qualitative interview | 8 people with PD | Self-management intervention | Assess feasibility | Improved knowledge and new strategies for living with PD and psychosocial benefits of meetings |
| **Multiple sclerosis (MS) – 36 studies** |  |  |  |  |  |
| Barlow, Turner, Edwards, and Gilchrist (2009) | RCT | 216 people with MS | Self-management (chronic disease self-management course, lay-led) | Determine effectiveness | Improved self-efficacy and physical health status |
| Barlow, Edwards, and Turner (2009) | Qualitative secondary analysis of Barlow et al. 2009 | 10 people with MS | Self-management | Explore experiences and value | Benefits perceived |
| Bombardier et al. (2008) | RCT (1 phone interview then 5 telephone counselling sessions) | 130 people with MS  | Telephone counselling for health promotion | Determine effectiveness | Improved health promotion behaviour, fatigue impact, mental health related QOL and self-reported physical activity |
| Cecil (2014)  | Case study description | 8 people with MS-Progressive | Support group (telephone) for home-bound patients (MS) | Consider first eight years of the group | Positive feedback |
| Dlugonski, Motl, and McAuley (2011)  | Repeated measures | 21 people with MS-RR | Self-management (internet) – increase physical activity | Examine efficacy using objective and self-report measures | Increased activity levels on objective and self-report measures  |
| Dlugonski, Motl, Mohr, and Sandroff (2012) | RCT | 45 people with MS-RR | Self-management (internet) – increase physical activity | Evaluate efficacy | Increased physical activity |
| Ehde et al. (2015)  | Single centre RCT + 6 and 12 month follow up | 163 people with MS | Self-management intervention (telephone delivered) | Evaluate efficacy | Significantly improved symptoms of fatigue, pain, depression, self-efficacy, activation, health related QOL, resilience, and affect.  |
| Feys et al. (2013) | Uncontrolled interventional study withrepeated measures | 42 people with MS | Education day with theoretical and practical sessions to increase physical activity level | Evaluate effects | Enhanced physicalactivity behaviour in persons without perceived disability |
| Finlayson, Garcia, and Preissner (2008)  | Qualitative | 28 people with MS, 3 healthcare practitioners  | Energy conservation (teleconference delivered) education program | Evaluation | Positive feedback but also limitations highlighted |
| Finlayson, Preissner, Cho, and Plow (2011) | RCT | 190 people with MS | Energy conservation (teleconference-delivered fatigue management program) | Explore effectiveness and efficacy | Improved fatigue impact and aspects of HRQOL |
| Finlayson (2005) | Pilot repeated measures | 190 people with MS | Energy conservation (group, teleconference) | Explore efficacy | Reduced fatigue impact and severity, better bodily pain and general health |
| Ghahari, Packer, and Passmore (2009) | Single group pre- and post-training comparisons | 9 people with MS and 2 people with PD (no separate analysis) | Online fatigue self-management program | Develop program and evaluate effectiveness | Decreased fatigue |
| Knoop, van Kessel, and Moss-Morris (2012) | Secondary analysis of RCT (van Kessel et al. 2008) | 70 people with MS | CBT- face to face and telephone for fatigue management | Assess mediators of effectiveness | Changing negative representations of fatigue crucial. CBT better than relaxation training (RT) |
| Mathiowetz, Matuska, and Murphy (2001) | Repeated measures- AB design | 54 people with MS | Energy conservation course on fatigue impact, self-efficacy and QOL | Evaluate effectiveness | Improved fatigue impact, self-efficacy and aspects of QOL |
| Mathiowetz, Finlayson, Matuska, Chen, and Luo (2005) | RCT with immediate - delayed control group cross-over design | 169 people with MS  | Energy conservation course for people with MS, some with mild cognitive impairment  | Assess the short-term efficacy and effectiveness | Decreased fatigue, increased self-efficacy and some aspects of quality of life |
| Mathiowetz, Matuska, Finlayson, Luo, and Chen (2007) | Follow-up secondary data analysis (see Mathiowetz et al. 2005) | 169 people with MS | Energy conservation course | Report of 1-year follow-up analysis | Decreased fatigue and improved quality of life maintained up to 1 year post-intervention |
| Miller et al. (2011) | RCT | 206 people with MS  | Self-management prompts (web-based, participant generated) | Investigate effectiveness & feasibility | No group differences in favour of self-management prompts |
| Moss-Morris et al. (2012) | Pilot RCT | 40 people with MS | Self-management (internet-based CBT self-management for MS, focus on fatigue)  | Feasibility | Reduction in fatigue severity and impact. Improved anxiety and depression and increased QALYs |
| Motl and Dlugonski (2011) | Repeated measures (interrupted time series) | 18 people with MS-RR | Self-management (internet) – increase physical activity | Examine effectiveness | Increased self-reported and objectively measured physical activity (small) |
| Motl, Dlugonski, Wójcicki, McAuley, and Mohr (2011) | Pilot RCT | 54 people with MS-RR | Self-management (internet) – focus on physical activity | Examine effectiveness | Increased self-reported physical activity levels  |
| Motl, Dlugonski, Pilutti, and Klaren (2015)  | RCT, pre- and post- intervention assessment (Secondary analysis) | 82 people with MS-RR | Behavioural intervention with goal-setting component to increase physical activity  | Examine efficacy | Participants in the behavioural intervention group had significantly higher physical activity levels than controls. The efficacy of the intervention was moderated by MS type, disability status, and weight status. |
| Mulligan, Treharne, Hale, and Smith (2013) | Qualitative analysis of the feasibility trial data  | 27 people with MS | Combined self-help and professional help “Blue Prescription” to minimize physical and social barriers to participation | Establish content and resources required for delivery of the approach | Increased participationin community-based physical activity physical activity of their choice |
| Navipour et al. (2006) | Pre- and post-intervention comparisons | 34 people with MS | Short-term self-managed graded exercise programme for various MS symptoms | Evaluates effects | Improved self-esteem and fatigue |
| Ng et al. (2013) | Pre- and post-intervention comparisons and follow-up assessment | 129 people with MS | Wellness program of psychological and physiologicalevaluations, lectures and workshops | Determine efficacy | Improved self-efficacy and self-perceivedhealth related outcomes, but no increase in physical activity |
| O'Hara, Cadbury, De Souza, and Ide (2002) | RCT | 169 people with MS | Self-management (group/ individual) | Test efficacy | Improved mental health, fatigue and perceived need for assistance |
| Pilutti, Dlugonski, Sandroff, Klaren, and Motl (2014) | RCTSecondary analysis of data from RCT (see Sandroff et al., 2014) | 82 people with MS-RR | Self-management (internet) - physical activity  | Examine effectiveness on secondary outcomes  | Improved fatigue severity and impact, anxiety and depression and physical activity level |
| Plow, Bethoux, McDaniel, McGlynn, and Marcus (2014)  | Pilot RCT – randomly allocated two-group repeated measures design, delayed-treatment contact group was control | 30 people with MS-RR | Patient education (customised pamphlets) | Explore effectiveness | Increased stage of change, social support, use of behaviour strategies  |
| Plow, Bethoux, Mai, and Marcus (2014) | Additional secondary analysis of Plow et al 2014 (above) | 30 people with MS-RR | Patient education (customised pamphlets) | Additional analysis | Improved physical activity, symptom severity, walking  |
| Rietberg, van Wegen, Eyssen, and Kwakkel (2014)  | RCT, pre- and post- trial assessment + 24 week follow up | 48 people with MS | Multidisciplinary rehabilitation (received either physical therapy, OT, social work, or any combination of these) | Assess efficacy | No significant differences at 12 or 24 weeks follow up for fatigue, functional independence, impact on QOL or social participation |
| Sandroff et al. (2014) | Secondary analysis of RCT (Motl et al., 2011) | 82 people with MS-RR | Self-management (internet) – increase physical activity in inactive MS | Secondary outcomes | Improved walking and improved cognitive processing speed in mild MS |
| Shevil and Finlayson (2009)  | Process evaluation (qualitative) | 35 people with MS | Self-management (group) | Identify feasibility | Positive feedback related to group and course content |
| Shevil and Finlayson (2010)  | Repeated measures | 35 people with MS | Self-management | Pilot findings | Improved knowledge, strategy effectiveness and self-efficacy |
| Smith, Hale, Mulligan, and Treharne (2013) | Qualitative secondary analysis (see Mulligan et al. 2013)  | 27 people with MS | Combined self-help and professional help “Blue Prescription” | Investigate experienceof participants | Therapy seen as supportive, motivating and enabling |
| Stuifbergen, Becker, Blozis, Timmerman, and Kullberg (2003) | RCT | 57 people with MS | Self-management (group) | Determine efficacy | Improved self-efficacy and health behaviour at 8MFU. Immediate improvement in QOL |
| Stuifbergen, Becker, Timmerman, and Kullberg (2003) | Additional analysis of RCT | 57 people with MS  | Self-management (group) | Describe experience with goal attainment scaling | Increased goal attainment  |
| Twomey and Robinson (2010) | Qualitative analysis | 8 people with MS | Fatigue management programme | Examine the experience of participants | Experience described in positive terms |

1. **Support for carers**

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| **Authors & Date** | **Design** | **Sample** | **Intervention** | **Aims** | **Key findings** |
| Diehl, Mayer, Förstl, and Kurz (2003) | Single group post-intervention qualitative interview +6 month questionnaire follow-up | 8 carers (FTD) | Caregiver support group | (1) To provide information, advice, and support to caregivers, (2) to establish needs of caregivers, (3) to encourage mutual support and develop coping strategies, (4) to evaluate the intervention | Reduced caregiver burden, enhanced coping strategies, establish new friends, rated programme as beneficial |
| Dowling et al. (2014)  | Randomised control pilot study | 24 carers (FTD) | One-on-one caregiver positive emotion skill-building intervention  | Assess feasibility | Improved scores on measures of positive affect, negative affect, burden, and stress for intervention group. |
| McKinnon, O'Connor, Savage, Hodges, and Mioshi (2013)  | Between groups | 21 carers (FTD) | Caregiver support group | Qualitative evaluation of intervention | Increased functional responses to fictitious scenario |
| Mioshi et al. (2013) | Between groups | 21 carers (FTD) | Caregiver support group | Verify utility of the intervention  | Burden and reaction to behaviours reduced. No change in depression, stress and anxiety |
| O'Connell et al. (2014) | Post group feedback | 10 carers (atypical early-onset dementia including FTD) | Caregiver support group | Develop and evaluate a support group for rural spouses of individuals with dementia | Positive feedback |
| Dipple and Evans (1998)  | Qualitative - interview | 27 (mix of HD carers and people with HD) | Support group  | Describe the support network associated with the group | Infrequent contact outside group meetings |
| Shah et al. (2015)  | Single group pre- and post- intervention questionnaires and post- interview | 7 carers (PD) | Caregiver tele-support group | Assess feasibility | No significant findings, but scores on Geriatric Depression Scale decreased from 4.2 to 3.0 |

**Key:** CONDITIONS: FTD = frontotemporal dementia, SD = semantic dementia, HD = Huntington’s disease, PD = Parkinson’s disease, MND = motor neurone disease, MS = multiple sclerosis, PPA = primary progressive aphasia, LBD = dementia with Lewy bodies, PDD = Parkinson’s disease dementia, AD = Alzheimer’s disease, MSA = multiple system atrophy, MS-RR = multiple sclerosis remitting relapsing subtype. DESIGN: RCT = randomised controlled trial. INTERVENTION: CBT = cognitive behavioural therapy.

Records identified from other sources
(n=35)

Records identified through database searching
(n=19,187)

Records after duplicates removed
(n=18,457)

Studies included
(n=140)

MS (n=74); PDs (n=19); FTDs (n=27); HD (n=10); MND (n=10)

## Eligibility

## Included

## Screening

Full-text articles assessed for eligibility
(n=732)

Abstracts screened
(n=2,508)

## Identification

Abstracts excluded
(n=1,776)

Not about condition of interest (n=96)

Not about relevant interventions (n=1,680)

Records excluded after title screening
(n=15,949)

Full-text articles excluded
(n= 92)

Not about condition of interest (n=41)

Intervention not relevant or cognitively-impaired participants excluded (n=397)

Article unobtainable (n=154)

**Figure 1.** Flow chart illustrating the process of study selection