

# An exploration of the physical activity life cycle in Huntington's disease

Submitted in accordance with the requirements for the degree of  
Doctor of Philosophy (PhD)

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## Abstract

This study aimed to explore how living with Huntington's disease (HD) impacts on the experience of physical activity (PA) across the stages of the disease.

The research questions were:

- 1) What are the experiences of PA participation across the stages of HD?
- 2) How do the nuances of living with a neurodegenerative disease such as HD affect engagement in PA?

The focus group (FG) method was used to explore the breadth of experiences of PA in the context of HD. Participants of eight FGs across the UK included people across the spectrum of HD with varying degrees of symptom manifestation, caregivers (family members/formal caregivers), and healthcare professionals. Framework analysis method (Ritchie and Spencer 1994) was used for data analyses. The process involved five stages: familiarisation, coding, indexing, charting and mapping, and interpretation of data to develop key themes. A key part of the process was development of an analytical coding framework to use in the indexing (of the data) stage. The data itself and a theoretical model (self-regulation model) were both used to develop the framework. The literature review identified a lack of theoretically underpinned qualitative research in PA and HD, therefore the self-regulation model (SRM) (Levanthal et al. 1984) was selected to explore PA in HD. Components of the SRM were used to develop a priori 'index codes' of the framework. Open coding of transcripts was also used to develop 'index codes' of the framework.

The findings highlighted that over the life-span of the disease the needs and abilities of people with HD change, and this has implications such as coping responses and strategies for how PA is experienced and sustained in HD. Social context and environment are key aspects that require consideration for PA in HD, and this has implications such as consideration of familial or caregiver support and general public awareness of HD for development of management and research interventions. The SRM facilitated understanding of participant experiences, however, it appeared that self-regulation of activities becomes more collaborative with the caregiver as HD progresses. As such, a modified version of the SRM that incorporates the increasingly collaborative regulation of PA has been suggested and is presented for understanding PA in HD.

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# Contents

Declaration and statements .....	i
Abstract .....	ii
Acknowledgements.....	iii
Contents .....	iv
List of appendices.....	vii
List of figures .....	viii
List of tables .....	ix
List of abbreviations.....	x
1. Introduction.....	1
2. Background of the researcher.....	9
3. Literature review .....	13
3.1. Chapter overview .....	13
3.2 Search strategy .....	13
3.3. The biomedical perspective of Huntington's disease and consideration of its impact on physical activity participation.....	15
3.3.1. Epidemiology of Huntington's disease.....	17
3.3.2. Current management of Huntington's disease and potential for self-management of Huntington's disease through physical activity.....	17
3.3.3. The stages of Huntington's disease and potential impact on physical activity (PA) .....	21
3.4. Huntington's disease from the personal perspective.....	31
3.4.1. Characteristics and challenges of Huntington's disease.....	32
3.4.2. Experiences of healthcare services.....	35
3.4.3. The 'HD family' .....	35
3.4.4. Summary of 'Huntington's disease from the personal perspective' .....	38
3.5. Current literature exploring experiences of physical activity in Huntington's disease and other long term neurodegenerative conditions.....	38
3.5.1. Overview.....	38
3.5.2. Experiences of physical activity in Huntington's disease .....	40
3.5.3. Experiences of physical activity in other neurodegenerative conditions.....	45

3.5.4. Summary: current knowledge of experiences of physical activity in Huntington's disease and other neurodegenerative diseases .....	53
3.6. Application of theoretical models to explore experiences of Huntington's disease and other long term neurodegenerative conditions.....	54
3.7. Chapter summary.....	70
4. Methodology .....	72
4.1 The researcher's perspective; 'my perspective' .....	72
4.2 Research design and rationale .....	73
4.3 Focus groups .....	74
4.3.1. Development of the focus group schedule .....	75
4.3.2. Participants and recruitment .....	76
4.4 Data collection.....	79
4.5. Data analysis.....	82
4.6. Ethical considerations .....	111
4.6.1. Ethical approval .....	111
4.6.2. Informed consent .....	111
4.6.3. Data management .....	112
4.6.4. Confidentiality and anonymity .....	112
5. Results and discussion of key themes .....	113
5.1 Chapter overview .....	113
5.2. The Focus groups .....	113
5.3. Introduction to the key themes.....	117
5.4 Key theme 1: The evolving representations of Huntington's disease and their impact on physical activity .....	121
5.5. Key theme 2: The varying social environment of the person with Huntington's disease and the impact on physical activity .....	149
5.6. Key theme 3: Achieving physical activity participation while coping with the nuances of Huntington's disease.....	169
5.7 Discussion: drawing together the key themes.....	186
6. Research conclusions and implications .....	198
6.1 Original contribution .....	198

6.2 Limitations .....	200
6.3 Implications for research and clinical implications.....	203
6.4 Final conclusions.....	213
References .....	215
Appendices .....	229

## List of appendices

- Appendix 1: Summaries of studies exploring treatment approaches in Huntington's disease
- Appendix 2: Summaries of studies of experiences of the personal experiences of living with Huntington's disease
- Appendix 3: CASP summary table: critical appraisal of studies related to experiences of Physical Activity in Huntington's disease
- Appendix 4: Summaries of studies of experiences of physical activity in Huntington's disease
- Appendix 5: Summaries of studies of experiences of physical activity (PA) in other neurodegenerative conditions
- Appendix 6: Summaries of studies from the literature review that used theoretical models to underpin the research
- Appendix 7: ENGAGE-HD study
- Appendix 8: Example of a flyer sent out to Huntington's disease Association Specialist HD advisors to circulate
- Appendix 9: Participant Information Sheet
- Appendix 10: Consent form
- Appendix 11: Research ethics committee approval letter
- Appendix 12: Detailed development of the focus group schedule
- Appendix 13: Focus group schedule and workshop schedule
- Appendix 14: Demographic questionnaire for focus group participants with Huntington's disease
- Appendix 15: Demographic questionnaire for focus group participants (caregivers)
- Appendix 16: Focus Group details
- Appendix 17: The analytical framework in NVivo
- Appendix 18: Extract of testing of the analytical framework by one of the EHDN fellows
- Appendix 19: Initial codes identified from open coding
- Appendix 20: Example of an annotation linked to data whilst the researcher was indexing using the analytical framework
- Appendix 21: Memo: 'Identity'
- Appendix 22: 'Physio Works' document; an evidence based document for the Chartered Society of Physiotherapy
- Appendix 23: Researcher's interpretation of theoretical development of the ENGAGE-HD intervention

## List of figures

- Figure 1: MRC framework for developing complex interventions 2008
- Figure 2: Map of literature review
- Figure 3: World Health Organisation International Classification of Disability and Health Model (WHO-ICF, 2001)
- Figure 4: Key constructs of Social Cognitive Theory (adapted from Bandura 1986)
- Figure 5: The Self-regulation Model (adapted from Levanthal et al. 1984)
- Figure 6: Example of developing themes adapted from Saldana (2009)
- Figure 7: Comparison of terminology for analytical framework components (adapted from Spencer and Ritchie, 1994; Gale et al. 2013).
- Figure 8: Example showing how one of the categories of the analytical framework was developed from open coding.
- Figure 9: Grouping of final codes to form the categories of the analytical framework from open coding
- Figure 10: Iterations of the names of one of the key theme and its subthemes
- Figure 11: Memo relating to development of key theme related to social environment
- Figure 12: Development of a key theme; spider diagrams and memos
- Figure 13: Illustration of how subthemes for this key theme emerged from the data indexed to the categories and codes of the analytical framework.
- Figure 14: Diagrammatic Representation of key themes and subthemes
- Figure 15: Key themes in relation to the context of physical activity participation in HD and the interwoven thread of 'change over time' and self/collaborative regulation.
- Figure 16: Illustration of the key themes mapped onto the Self-regulation model (Levanthal et al. 1984)
- Figure 17: The proposed adapted self-regulation model specific to physical activity in HD
- Figure 18: Determinants of PA behaviour and ENGAGE-HD intervention components

## List of tables

- Table 1: Total Functional Capacity (TFC) stages of disease
- Table 2: The a priori codes developed from the Self-regulation model
- Table 3: Refined codes and final refined codes of the analytical framework identified through open coding
- Table 4: Working analytical framework
- Table 5: The final analytical framework
- Table 6: Indexed quotes and relevant codes/categories of the analytical framework
- Table 7: Example of heading in coding matrix
- Table 8: Example of some quotes indexed to codes and categories of the analytical framework that related to the concept of the 'social environment' as an initial theme.
- Table 9: Quotes indexed to categories of consequences and timeline
- Table 10: Focus Group details
- Table 11: Examples of some initial codes from open coding and quotes linked to them

## List of abbreviations

AD	Alzheimer's disease
CASP	Critical Appraisal Skills Programme (Oxford UK)
COMMET-HD	Community-based Exercise Therapies for Huntington's Disease
EHDN	European Huntington's Disease Network
FG	Focus group
HD	Huntington's disease
HDA	Huntington's Disease Association
MS	Multiple Sclerosis
SHDA	Specialist Huntington's Disease Advisor
PA	Physical activity
PD	Parkinson's disease
RA	Rheumatoid arthritis
PEO	Population Exposure/condition Outcome/theme
PICO	Population Intervention Comparison Outcome
RCT	Randomised controlled trial
SCT	Social cognitive theory
SRM	Self-regulation Model
TMSC	Transactional Model of Stress and Coping
WHO-ICF	World Health Organisation International Classification of Disability and Health
FA	Framework analysis
WHO	World Health Organisation

## 1. Introduction

Huntington's disease (HD) is a progressive neurodegenerative disorder caused by a genetic mutation that follows an autosomal-dominant pattern of inheritance; each affected person has a 50 per cent chance of transmitting the disorder regardless of sex (Harper 2005). Following clinical diagnosis, over approximately ten to fifteen years, progression of cell death in the brain causes increasingly severe physical, behavioural and cognitive impairments. Apathy is a common characteristic from early on in the disease process, and the movement disorder is the most striking visually perceptible consequence of HD. Chorea, impaired balance and gait cause an altered way of walking. People with HD have been described as looking 'slightly drunk', which also speaks to the social stigma that HD families may experience (Roos 2010; Wexler 2010).

In brief, from early to later stages of HD the needs and abilities of people with HD constantly change. Symptoms cause individuals to experience increased difficulties in functioning in all aspects of everyday life (Walker 2007; Naarding et al. 2009; Tabrizi et al. 2013). This may include the inability to maintain employment, difficulties in organising and planning, keeping appointments, impaired mobility, increased difficulties in washing and dressing independently, cooking meals, impaired speech and communication, impaired swallow, inability to dual-task, difficulty maintaining roles within the family and engaging in health behaviours such as physical activity (PA) (Downing et al. 2010). Disease duration, motor and or cognitive impairments and psychological concomitants including the way in which individuals cope with HD (Helder et al. 2001) compromise physical and psychosocial well-being of people with HD.

The heterogeneous nature of HD and the different trajectories of progression within different individuals is a further challenge (Rosas et al. 2008). For example, somebody with HD who has significant behavioural and cognitive symptoms may lead a chaotic life for a number of years possibly requiring input from psychiatric services, social services, coming into contact with legal and judicial services, and possibly isolating themselves from family and friends. Others may have little or no insight into the symptoms caused by HD or be in denial, which leads to inappropriate decisions and behaviours. Alternatively, an individual may have little or no problems cognitively or behaviourally but suffer severe movement disorders including chorea and dystonia.

The devastating impact of HD has been referred to as a “spectre” hanging over families living in its shadow (Maxted et al. 2014). As onset is usually around middle age, relationships and roles within the family unit change (Williams et al. 2007; Downing et al. 2010; Maxted et al. 2014). It is not uncommon for family members to be caring for more than one individual with HD at the same time while younger generations are at risk of having inherited the mutated gene. Caregivers play an integral part in the daily lives of people with HD as the disease progresses. As such, caregivers have much responsibility for activities of daily living including functional tasks, social activities and healthcare appointments (Downing et al. 2010). However, the stigma associated with such a genetic disease can also be divisive within families (Wexler 2010) and cause isolation from society in general, compromising quality of life and wellbeing for people with HD.

Despite the gene discovery more than twenty years ago and ongoing international efforts to identify possible targets and interventions (Johnson & Davidson 2010) there is currently no definitive treatment to prevent, delay or slow the disease process (Novak & Tabrizi 2010). Current symptomatic management of HD relies on pharmacological approaches to target chorea, rigidity, depression, irritability and altered sleep cycles (Phillips et al. 2008). It is clear that there is further significant work needed in developing effective curative and symptomatic treatment. There is now increased recognition of the potential of non-pharmacological approaches in the management of HD.

The role of the multidisciplinary team has been emphasised regarding long term management including physiotherapists, social workers, general practitioners, neurologists, geneticists, psychiatrists, occupational therapists, speech and language therapists, dieticians, community mental health teams, and social workers (Novak & Tabrizi 2010). Experiences of healthcare service support are varied though, and provision is inconsistent; gaining access to appropriate healthcare services is reported as challenging for people with HD and their caregivers (Etchegary 2011). This has been attributed to healthcare professionals’ lacking understanding of HD, insufficient specialist services and poor integration between health and social services (Aubeeluck et al. 2012; Skirton et al. 2010; Maxted et al. 2014; Welsh Government 2014). Due to the longevity and lack of cure for HD, long term management of the condition is required. Support for self-management approaches in neurological conditions which reduce the need for ongoing rehabilitation has been a focus of government drivers for practice in recent years (Welsh Government 2014; Department of Health 2015; Department of Health 2016). Empowering patients to self-manage their condition and

decisions about care has been recognised as a way of better meeting patient's needs, improving quality of life and reducing burden on the health care system (Wales Audit Office 2014; Department of Health 2015; McKenna et al. 2015; Department of Health 2016). The emphasis to strive for better patient self-management is seen in the NHS Outcomes Framework for 2016-17. One aim of the five domains addresses 'Enhancing quality of life for people with long-term conditions' (Department of Health 2016). The framework suggests that ensuring people feel supported to manage their condition and improving quality of life and functional could help achieve this (Department of Health 2016). Specifically, in Wales, and in neurological conditions the 'Together for Health - A Neurological Conditions Delivery Plan' (Welsh Government 2014) sets out government expectations for care and services. Support to self-manage where appropriate is a delivery expectation under the 'Living with a neurological condition' theme of the delivery plan. The 'Bridges programme' is an example of an approach that supports self-management in stroke (Jones et al. 2009). The programme which is underpinned by social cognition theory (SCT) takes a holistic approach to encouraging self-management.

The importance in the use of theory to underpin such resources or interventions has been recognised; a criticism of previous interventions has been that they lack underpinning theory (Jones et al. 2009). In developing complex interventions, the value of theory is recognised in the focus that it can give without being restrictive, and enhance understanding of the phenomena of interest. Use of theory can also improve intervention quality, through identifying necessary concepts or elements for inclusion (Green 2000). Just one example of a commonly applied framework is the 'behaviour change wheel' (BCW); a framework for use in developing behaviour change interventions and has been applied to tobacco control, obesity and stroke (Michie et al. 2011; Connell et al. 2015). The BCW was developed by Michie et al. (2011) following an evaluation of existing frameworks for behaviour change, to meet criteria of coherence, comprehensiveness, and a clear link to an overarching model of behaviour. The framework suggests that for behaviour change to occur, certain needs or criteria need to be met. These are capability (physical or psychological ability to enact the behaviour), motivation (reflective and automatic mechanisms that activate or inhibit behaviour), and opportunity (physical and social environment that enables the behaviour). In 'Bridges' for example, 'setting personal goals which may include PA' is incorporated, and goal setting is a component of SCT. As such the use of theory in developing the 'Bridges' intervention has been helpful in developing a programme that people engage with because it is meaningful to them (McKenna et al. 2015).

Engaging in meaningful activities is also important in HD where quality of life both in terms of physical and psychosocial well-being, are severely affected (Helder et al. 2001). Supportive resources to engage in PA are important and relevant, especially if robustly underpinned by appropriate theory. Furthermore, specifically in HD, there exists a growing body of evidence that highlights the potential that PA may have in symptom management, improved quality of life and disease modification (van Dellen et al. 2008; Renoir et al. 2012; Busse et al. 2013; Quinn et al. 2016). A number of exercise programmes delivered as part of small scale clinical feasibility studies were feasible and safe, and effect estimates suggested some benefit such as improved strength, health related quality of life and achievement of functional goals in HD (Khalil et al. 2012; Busse et al. 2013; Quinn et al. 2014). Effects of exercise have been found to positively influence issues associated with HD (Grimbergen et al. 2008; Busse et al. 2009; Quinn et al. 2014).

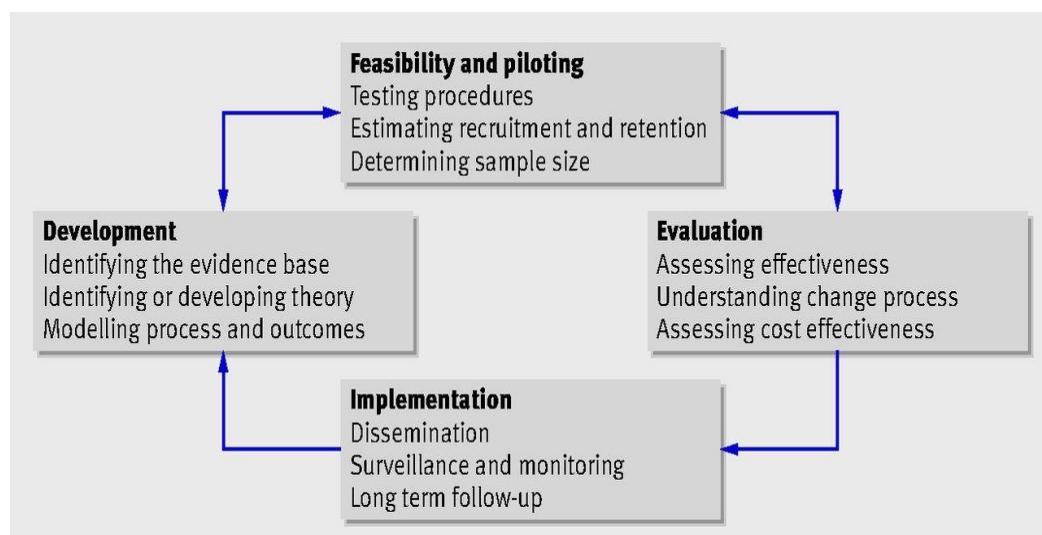
Positive influences of PA have been observed specifically regarding activities balance confidence, fatigue, PA levels, functional measures, measures of gait and health related quality of life in Parkinson's Disease (PD) and Multiple Sclerosis (MS) (Miyai et al. 2002; Caglar et al. 2005; Ashburn et al. 2007; Kargarfard et al. 2012; Learmonth et al. 2013; Schenkman et al. 2012; Guidi et al. 2013; Gervasoni et al. 2014). PA interventions are complex in terms of the interacting components of the intervention (context, participant, person(s) delivering the intervention, the intervention itself). Guidance published in 2000 and reviewed in 2008 by the Medical Research Council (MRC) incorporated a framework for developing and evaluating complex interventions. Guidance for developing process evaluations was published in 2015 (Campbell 2000; Craig et al. 2008; Moore et al. 2015). The MRC framework in 2000 suggested that small scale feasibility studies should precede larger scale randomised controlled trials (RCTs).

The MRC framework was developed following the recognition of the need for a more defined approach to complex intervention development and implementation. The high number of negative trials of complex interventions being produced where the reasons for failure were not clear (Campbell 2000) needed to be addressed. Failure of the interventions could neither be attributed to the interventions' ineffectiveness or failure of other components such as delivery or intervention implementation because of a lack of transparent processes. The revised guidance in 2008 called for clear definitions of processes, where interventions were developed with transparency and the support of theory; using qualitative methods to support development and (where relevant) monitoring of complex interventions.

Complex interventions are those which contain several interacting components and may be delivered at a community, group or individual level. Complex interventions have been implemented in numerous areas, including the health service, public health practice and areas of social policy that affect health (e.g. education). Examples include behavioural interventions targeting health professionals' behaviour, and behavioural interventions to increase PA in different patient populations (Moore et al. 2015). Several dimensions of complexity exist in such interventions; the range of possible outcomes, their variability in the target population, behaviours that are required by participants receiving the interventions and the interactions between components within the interventions. For example, in delivering an educational intervention, there is a need to consider the development of educational materials, the skills of those delivering educational materials, the context in which they are delivered and the people that the intervention targets (their behaviour, social context). Indeed, there are numerous components which could affect delivery and intervention outcomes.

The guidance revision in 2008 was prompted because significant experience and knowledge had been gained since 2000 (Anderson 2008). It was also clear by 2008 that the original model required modification. The authors realised that non-pharmacological studies of complex interventions do not necessarily fit a linear model as had originally been developed in line with the sequential phases of drug development. The model was adapted (figure 1) and took into account that the process of developing complex interventions may be flexible in terms of the stages of development, piloting, evaluation and implementation, depicted by the multidirectional arrows in figure 1.

Figure 1: MRC framework for developing complex interventions (Anderson 2008)



Another consideration was that since publication in 2000, the conceptualisation that RCTs need to demonstrate fidelity through standardisation of components of the intervention had been challenged. The revised guidance discusses the importance of considering and understanding the context in which complex interventions are delivered and highlights the significance of the intervention-context interaction. The guidance places more emphasis on the need for standardisation through defined processes and function of the intervention, rather than of individual components within the intervention. Interventions that can be tailored to local conditions may be more effective (Hawe et al. 2004) and therefore understanding local conditions or ‘context’ is vital. In trying to understand context, qualitative approaches may be particularly useful because they allow the researcher to explore social or behavioural processes where quantitative methods are limited (Lewin et al. 2009).

The MRC framework emphasises the importance of qualitative research to develop better interventions and better design evaluations that could be successfully implemented in clinical practice and quality research (evaluations). The process evaluation guidance recognises that the interaction between participants and the intervention will be shaped by participant attitudes, beliefs and pre-existing circumstances. Viewed in this way, contextual factors can act as moderators of outcomes of interventions as well as implementation. This means that the same intervention in a different context may have different outcomes. The process evaluation guidance published recently suggests that quantitative and qualitative analyses can “build upon one another”; where qualitative data can be useful in explaining quantitative findings and quantitative data may be used to test hypotheses developed or generated by qualitative data (Moore et al. 2015).

The use of theory and qualitative research at numerous stages during the research process is suggested. Qualitative research can be used to explore implementation of complex interventions to monitor, help modify according to local context to ensure appropriateness to particular settings and support development and delivery of complex interventions. Qualitative enquiry has been used at different stages in the development, implementation and evaluation stages of complex interventions. However qualitative methods to help evaluate studies of complex healthcare interventions remain underused and poorly integrated (Lewin et al. 2009). This is certainly true for HD where current literature exploring the personal experiences of PA in HD is mainly limited to a number of qualitative studies ‘tagged’ onto investigations of specific exercise programmes as intervention process evaluations. Beyond the exercise research studies there is currently very little in terms of resources specifically

supporting people with HD to engage in PA. This is possibly because until recently there has been a lack of robust studies of exercise interventions to underpin an evidence base that demonstrates potential benefits of PA in HD specifically.

In relation to the MRC framework (figure 1), the evidence base for the potential benefit of regular exercise is becoming established and is growing (quantitative research) but the researcher recognised that gaining the crucial understanding of how HD impacts on participation in PA, (which could determine success or failure of an intervention), was not being addressed. Importantly, studies of PA in HD were focussed on specific interventions in specific contexts and there is lack of integration of theory and methods by which to understand participant experiences and why aspects of the exercise programmes might have been successful or unsuccessful. This is a recognised issue and has been identified by the Medical Research Council (MRC) in terms of improving quality of research developing ‘complex interventions’ such as PA or exercise interventions (Moore et al. 2015). In consideration of the MRC framework, quantitative research in HD and PA has been carried out and is being taken forward but there is a need for underpinning theory and knowledge to further develop and evaluate complex interventions, achievable through qualitative approaches. Furthermore, ‘implementation mapping’, describes how complex interventions can be developed, following a detailed six step process to capture intervention component development and to integrate the underlying theoretical foundations into a detailed logic map (Bartholomew et al. 2016). Indeed, a common limitation of the studies exploring PA in HD to date is a lack of strong qualitative work to support the quantitative aspects, except for one recent study (Busse et al. 2017). Methodological shortcomings of the studies of PA in HD include lack of theoretical underpinning, responses being subject to recall bias because of the length of time between receiving the intervention and collection of the experiential data, and having the same person delivering the exercise intervention and conducting the interviews (Zinzi et al. 2007; Quinn et al. 2010; Khalil et al. 2012).

Given the burden on the patients, families and healthcare services over an extended period of time, engaging in activities which may enhance quality of life and reduce health service burden is important. At present there is very little in the way of existing supportive resources for people with HD if they want support to become more active or initiate a PA routine. With apathy being a common characteristic of HD and the tendency for people with HD to be sedentary, (Busse et al. 2013) engagement in PA could be challenging.

It appears that the following are therefore vital; (1) Understanding of how the nuances of living with HD impact on physical activity across the spectrum of the disease, and (2) developing appropriately structured, theoretically underpinned interventions to support engagement in physical activity for people with HD. This research attempts to address these issues. The aim of this research is to ***explore how living with HD impacts on the experience of physical activity participation across the stages of the disease.***

A literature review was conducted to (1) explore the bodily symptoms and personal experiences of HD and consider how they may potentially impact on engagement and participation in physical activity, (2) develop an understanding of the existing literature related to the experiences of physical activity for people with Huntington's disease and identify the gaps in knowledge and (3) identify a relevant theoretical model to underpin this research.

The literature review informed development of the research questions which are presented at the end of the literature review chapter. The methodology chapter then provides description and rationale of the research methods used to address the research questions. Qualitative research approaches allow collection of experiential data and are therefore most appropriate to facilitate exploration of experiences and perceptions of people about a particular topic. The focus group method and a theoretically guided data analysis using an analytical coding framework were used to explore experiences and perceptions of people with HD and their caregivers in relation to how the nuances of HD impact on physical activity (PA) to improve understanding of PA in HD. The post-modern social constructivist perspective (Yilmaz 2007) was adopted to facilitate a focus that considered participant realities of their life worlds, as constructed by them and influenced by their experiences of HD in PA. Acknowledging that the existence of individuals is embedded within a complex set of social structures, the post-modern social constructivist perspective facilitates exploration of social, environmental and cultural influences (Yilmaz 2007). This could provide a richer insight into participant experiences of participation in PA rather than purely focussing on the bodily experiences of HD. It is key to acknowledge that this research has also been informed by the researcher's background and perspectives.

## 2. Background of the researcher

The researcher's (my) background as a physiotherapist and interest in neurological conditions and research led to me taking a role as a physiotherapy research assistant on a randomised controlled trial (RCT) of exercise in Huntington's disease (HD) (ISRCTN 59910670). The role involved trial coordination and delivery of a complex intervention (exercise intervention) in community gyms around South Wales with people with HD. The title of the RCT was "Community-based Exercise Therapies for Huntington's Disease" or "COMMET-HD". The RCT was based at Cardiff University and investigated the safety, feasibility, acceptability and effects of a supervised exercise intervention in HD. The interest and ideas for developing the research presented here emerged as a result of working closely with people with HD in supporting the participants in the exercise intervention and leading on the post intervention process analyses (Debono et al. 2012).

Delivering the intervention involved organising gym sessions, supervising and supporting individual participants in their local gym over 12 weeks. The intervention consisted of aerobic exercise on an exercise bike and strengthening exercises using weights. The goals of the sessions were progressed collaboratively with participants. Amongst other observations and experiences, good communication skills and pragmatism were required in terms of supporting participants to actually attend sessions as well as to do the exercises. Gaining understanding of the challenges faced by the individuals also helped with problem-solving to support the individuals during sessions. In delivering the exercise programme for the RCT it was clear to me that through informal conversation, observation during the sessions and recording reflective notes that for someone with HD, attending a gym session was not as straightforward as simply turning up and completing a work out. Participant experiences were varied and numerous issues were encountered. Examples included forgetting the appointment for the session, something else occurring last minute related to HD and taking priority, incidents happening while on their way to the gym, the need for carer support, having to deal with logistical issues such as arranging taxis as participants could not do it themselves. These were all challenges that had to be dealt with by myself whilst delivering the intervention.

In seeing how people with HD participated in the exercise programme, I recognised the value of the knowledge that could be gained from the participant experiences in terms of how the nuances of HD affected their ability to participate. Furthermore, none of this rich insight into the experiential side of the intervention was being formally captured. When delivering the intervention, a keen interest in how people with HD experience

participation in PA and how living with HD impacts on their experiences developed. It became clear to me that reduced PA could be expected given the impact of the cognitive, behavioural and motor symptoms of HD. Some ‘barriers’ that were witnessed in delivering the exercise intervention have been previously identified in PA in neurological conditions including difficulties with transport (Elsworth et al. 2011). However, the complex nature of HD (the combination of physical, cognitive and behavioural impairments) which makes it good as a model for other neurodegenerative diseases, makes it more complex by comparison. In supporting people who face a combination of cognitive, behavioural and physical challenges, (even over a short time period), I realised that there is more to learn because of the complexities caused by HD that are not seen in combination in other diseases. Considering that the people supported during the exercise programme were deemed to be at the same stage of HD, there was a plethora of different experiences and different strategies used with different people in communication, organisation, supervision and assistance with equipment. Reflecting upon this, it became clear that there must be much more to be learned from people living with HD at different stages about what their symptoms mean for engaging in PA.

Plainly visible symptoms including the movement disorder and impaired gait were the subject of embarrassment for some COMMET-HD intervention group participants. Embarrassment and lack of confidence in entering a gym environment may be barriers that people without medical conditions may experience. However, visible impairments caused by HD clearly identifies somebody as being different, ill or disabled, or to some outsiders unaware of HD, the person with HD appears inebriated. This is something which I have witnessed and been told by HD families, and was a problem for one of the participants. The stigma of symbols representing disability and the assumption that people with movement disorders are inebriated have been reported in other neurological conditions such as MS (Dennison et al. 2011).

For some, having their spousal caregivers present was helpful in terms of helping build their confidence in what they were doing and being in the gym environment. Having witnessed the reactions of others in the gyms where people would take a lot more than a short glance at the participants, it became transparent as to why participants might be self-conscious and or embarrassed. Feeling self-conscious was something that I had dealt with at times during those sessions and thoughts such as ‘what is the best way to deal with this unwanted attention?’ would surface. The behaviour of others posed an issue, which in a healthcare environment might not occur because people’s expectations of what they see may differ. In a gym environment the expectation is to

see 'normal', 'healthy' people, not those with movement disorders, who to the unaware, may appear to be inebriated (Dennison et al. 2011). This dilemma could apply to other health care professionals working in the community with HD. These social experiences may have implications for how people with HD go about their daily life and participation in PA and therefore for complex intervention development.

Although there was a strict protocol to follow regarding the exercise session content at the gym, there were additional components required to facilitate the gym sessions and to try to make the sessions a positive, successful experience for the participants. The argument that tailoring interventions to local context is something that was found to be useful in maintaining adherence (Hawe et al. 2004; Craig et al. 2008). For example, being flexible with the timing of sessions or being pragmatic about a particular exercise technique to ensure safety. Much insight was gained from experiencing and being a part of the participant experiences. The importance of understanding and capturing those experiences became apparent to me in terms of using that knowledge to better support people with HD in being active. What this also highlighted was that the understanding gained could have been very informative for development of future trials investigating exercise in HD. The qualitative aspect of the COMMET-HD RCT was designed to evaluate the exercise intervention safety, acceptability and feasibility. The thematic analysis of the process interviews was carried out by myself. On reading the transcripts, the depth of the data did not reflect the rich insights I had gained. This was partly I believe due to numerous one word answers to questions which was perhaps a limitation of the individual interviews. Not all participants had family members and or caregivers present during the interviews which may have been a limitation.

Family members and, or caregivers may have prompted them to elaborate on their answers with more success than the interviewer or created a different dynamic that attenuated any 'power differences' between interviewer and interviewee (Karnieli-Miller et al. 2009). This may have helped create an environment conducive to more equal participation of the interviewer/interviewee during the interview, eliciting more detailed responses. In addition, because of the potential cognitive impairments of the participants, caregivers could have helped them to remember or identify issues that they did not pick up on (Carlozzi and Tulsky 2013). Another argument for including the caregiver perspective is that they have a potential role in supporting and encouraging PA participation. The caregiver or family member can influence the person's daily activity and therefore their own beliefs and attitudes towards exercise can impact on PA participation of the person they live with and possibly help to care for. In consideration of this, focus groups may have been more beneficial, which create a

different type of dynamic where the expectation is not on one individual to respond to questions. Although not used in COMMET-HD, the limited formal research with caregivers could support the future use of focus groups with people with HD and their caregivers (Carlozzi and Tulsky 2013). Another interview limitation was the narrow focus of the questions. As such, being focused primarily only on certain aspects (of safety, acceptability and feasibility), the evaluation missed out on capturing all of the rich experiential data that could have built on the findings of the trial to be even more informative. Indeed, capturing such experiential information could help understanding of mechanisms which have a bearing on whether complex interventions such as that exercise programme are successful.

On reflection, although the RCT incorporated process evaluation in terms of interviews post intervention, the qualitative inquiry was not of sufficient depth. More in depth process evaluation could have shed light on important factors with regard to implementation of the intervention, mechanisms of action and how the context in which the intervention was delivered affected implementation and outcomes (Moore et al. 2015). The team were going through a learning process in terms of how to use the new MRC guidance for developing complex interventions and adapt the approach to design of RCTs. Furthermore, although the trial protocol incorporated the use of process evaluation, the detailed guidance for process evaluation which is available now, was not yet in existence then (Moore et al. 2015).

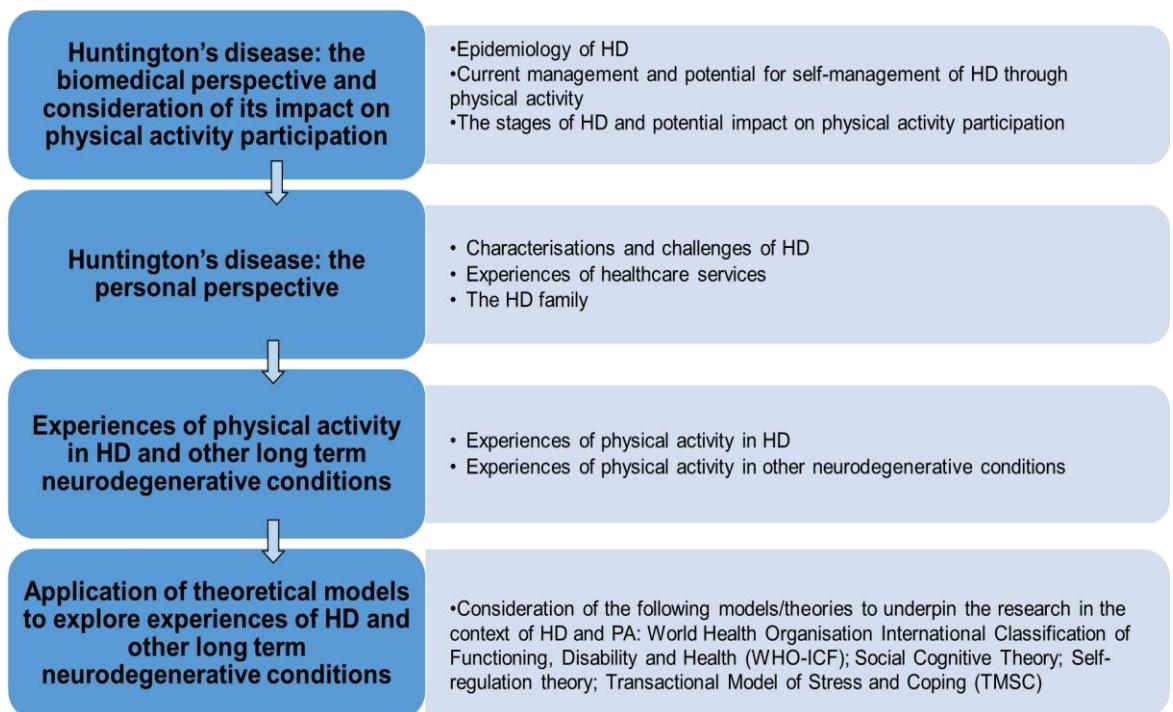
The foundational ideas for conducting the research presented here developed from the experiences described above and my passion to build on the work that was being developed by the team of researchers that I worked with. Whilst the research team focused on developing the quantitative evidence base for exercise in HD, I wanted to explore the experiential aspects in more depth and utilise a different method for data collection that may yield richer data, avoiding the limitations associated with the individual interviews of the COMMET-HD study. As described in the introduction chapter, this could be informative to design of future exercise interventions in research and clinical practice to support people with HD to self-manage through PA.

### 3. Literature review

#### 3.1. Chapter overview

A literature review was conducted to (1) explore the bodily symptoms and personal experiences of Huntington's Disease (HD) and consider how they may potentially impact on engagement and participation in physical activity, (2) develop an understanding of the existing literature related to the experiences of physical activity for people with Huntington's disease and identify gaps in knowledge and (3) identify a relevant theoretical model to underpin this research. The map in figure 2 may be useful for literature review navigation. Working from top to bottom, the figure illustrates the main headings of the literature review sections in order (darker blue) with the subheadings found within the main sections to the right of each one (in light blue).

Figure 2: Map of literature review



#### 3.2 Search strategy

Database searches of ASSIA, Pubmed and (Allied and Complementary Medicine), Embase 1996 to 2016 February 29, Ovid MEDLINE(R) without Revisions 1996 to February Week 4 2016, PsycINFO 2000 to February Week 4 2014, PsycArticles Full Text, Pubmed and Applied Social Sciences Index and Abstracts (ASSIA) were carried out to identify relevant research for literature review section. Limits were placed on the searches including 'full text', 'English language' and date limit of the year '2000' to

current. Date limits were initially set to within 10 years (2014 to 2004) but due to the dearth of literature in HD this was extended to the year 2000 onwards as there seemed to be a cluster of work developed from around this time. When the limits were changed to include the year 2000, six more relevant papers were identified. The main literature search was carried out in 2014, this was updated throughout the rest of the candidature, with the final search conducted in 2017. Papers identified since 2014 include recently published randomised controlled trials of exercise and intervention development papers in HD (Busse et al. 2016; Busse et al. 2017).

Search strategies to identify relevant literature were developed using the 'PICO' and 'PEO' approaches (Bettany-Saltikov 2012) PICO is a model designed to help develop a useful search strategy using the following: P= patient/population/problem; I = Intervention; C = Comparison; O = Outcome. PICO is helpful in identifying quantitative studies. PEO is a model to facilitate searching of qualitative literature: 'P'=patient/population; 'E'=exposure/condition; 'O' = outcome/theme (Bettany-Saltikov 2012). Both approaches were used and the search terms used are listed in each relevant literature review section.

Using both PICO and PEO aided exploration of the biomedical aspects of HD in terms of symptom presentation and epidemiology and experiences of living with HD allows the researcher to build a holistic picture of the disease. Being able to engage in PA is key to achieving the potential benefits for people with HD. Understanding HD may help understanding of how it could affect PA participation. The researcher explored this reflexively in terms of current knowledge of HD in the section 'Potential impact of HD on physical activity'. A review the current literature regarding experiences of HD and PA then follows, which was facilitated by the PEO approach. Reviewing literature about experiences of PA in HD allowed the researcher to establish the current knowledge base and identify gaps. In addition, research in other neurodegenerative diseases was considered because of the lack of current qualitative evidence about PA in HD. A key part of developing the research proposal was the identification of a theoretical model to underpin the research. Theoretical models and theories that had been used within the literature regarding experiences of HD and experiences of PA in other neurodegenerative conditions (due to lack of any models used in the HD literature regarding PA) were considered to identify one that would be relevant in the context of HD that could be utilised to develop the design of this research.

The Critical Appraisal Skills Programme (CASP) was used to appraise the literature (CASP 2014). Facilitated by CASP, the review focussed on quality of reporting,

methodological rigour and conceptual depth and breadth of studies as suggested by Hannes (2011). The relevant CASP qualitative and quantitative CASP tools were used to appraise the qualitative studies and quantitative studies.

### **3.3. The biomedical perspective of Huntington's disease and consideration of its impact on physical activity participation**

The PICO approach was followed to identify literature that facilitated understanding and helped the researcher describe biomedical aspects of HD found through observational, prospective and retrospective studies. Key search terms for Population (P) were 'Huntington's disease', 'Huntington's chorea'. Key terms for Intervention (I) and Comparison (C) were not used as observational studies were the main focus of this search to help understanding of HD. Outcome (O) key words included 'symptoms', 'pathology', 'impairments', 'pre-manifest', 'prodromal', 'prodrome', 'early stage', 'mid stage', 'late stage'. Limits placed on the searches included full text only, English language.

Huntington's disease (HD) is a neurodegenerative genetic disease that causes physical deterioration, cognitive impairment and behavioural / psychiatric problems. The HD gene (IT15) was identified on the short arm of chromosome 4 (4p16.3) in 1983 using DNA markers. In 1993, the HD gene was isolated and the mutation that causes HD was discovered, which is an extended DNA trinucleotide of cytosine – adenine – guanine amino acids (CAG) in the Huntington gene. The extended CAG repeat creates an abnormally long polyglutamine repeat in the huntingtin protein (Ross & Tabrizi 2011). This results in a toxic gain of function to the detriment of cellular function (Walker 2007). HD gene expression is ubiquitous and found in peripheral tissues but predominantly affects specific areas of the central nervous system (CNS) (Sharp et al. 1995).

HD pathogenesis causes selective changes in the brain. Prominent cell loss and atrophy occurs in the caudate nuclei, putamen (of the basal ganglia which make up the striatum) and particularly the medium spiny neurones. Specific neuronal population loss is suggested to account for the distinct phenotype of motor, behavioural and cognitive symptoms of HD (Imarisio et al. 2008). Onset is usually between the age of 35 and 60 (Walker 2007). Symptom onset depends on genetic mutation length of the Huntington gene but environmental modifiers including social environment and lifestyle are also thought to be influential (Georgiou et al. 1999; Anca et al. 2004; van Dellen &

Hannan 2004). Before formal diagnosis, but with confirmation of a genetic test, people are said to be gene positive or at prodromal stage.

HD progresses over fifteen to twenty years following manifestation of clinically observable symptoms. Following diagnosis, the disease progresses from early stage, to mid-stage, to late-stage and finally end stage (discussed in more detail in 'stages of HD and potential impact on physical activity'). Although stages are defined, in reality there is blurring between stages as people progress; an individual may be considered early to mid-stage. The Unified Huntington's disease Rating Scale Total Functional Capacity (UHDRS TFC) (table 1) indicates an individual's general function. TFC provides a general guide to disease stage by assessing capacity to carry out functional tasks of daily living (within five domains) (Shoulson & Fahn 1979). Higher scores for each domain indicate better function. Stage I (early) refers to TFC 11-13, stage II (middle) refers to TFC 7-10, (mid) stage III refers to TFC 3-6, Stage IV (late) refers to TFC 1-2 and stage V (late/end) refers to TFC 0.

In consideration of the World Health Organisation International Classification of Functioning and Disability (World Health Organisation 2001), examples of reduced participation, impairment in body structure and function and restricted activities are common. Progression of HD leads to complete loss of independence and function. Death usually occurs due to secondary causes such as complications of falls, heart disease, nutritional deficiencies or aspiration (Walker 2007).

Table 1: Total Functional Capacity (TFC) stages of disease

Engagement in occupation		Capacity to handle financial affairs		Capacity to manage domestic responsibilities		Capacity to perform activities of daily living		Care can be provided at home	
Description	Score	Description	Score	Description	Score	Description	Score	Description	Score
Normal	3	Normal	3	Normal	2	Normal	3	Home	2
Reduced capacity for usual job	2	Requires slight assistance	2	Impaired	1	Minimal impairment	2	Home or chronic care	1
Marginal work only	1	Requires major assistance	1	Unable	0	Gross tasks only	1	Full time skilled nursing	0
Unable	0	Unable	0			Total care	0		

### **3.3.1. Epidemiology of Huntington's disease**

The incidence of HD in the UK has been estimated at 7.2 per million person-years (Wexler et al. 2016). Similarities in numbers have been reported between males and females (Evans et al. 2013). Exceptions where HD is concentrated in particular areas include Tasmania and Lake Maracaibo, Venezuela (Wexler et al. 2004). UK prevalence, which could be an underestimate because of non-disclosure or non-diagnosis means that HD is classified as 'rare' under European Commission criteria. The criteria for 'rare' is where less than five per 10,000 people are affected (Rath & Kelly 2016). This statistic is irrelevant to the families affected by HD who live with the condition day in day out for many years. The genetic nature means that many generations of one family may suffer the enduring consequences of HD. A striking example is of Lake Maracaibo in Venezuela where most of the 18,149 people with HD from ten generations can be genetically linked to one woman (Wexler et al. 2004).

### **3.3.2. Current management of Huntington's disease and potential for self-management of Huntington's disease through physical activity**

Key search terms used for Population (P) were 'Huntington's disease', 'Huntington's chorea'. Key terms for Intervention (I) were 'treatment', 'management', 'medication', 'rehabilitation', 'physiotherapy', 'multidisciplinary', 'medical management'. Outcome (O) key words included 'symptoms', 'pathology', 'impairments', 'pre-manifest', 'prodromal', 'prodrome', 'early stage', 'mid stage', 'late stage'. Limits placed on the searches included full text only, English language. Intervention studies identified related to different exercise interventions (see appendix 1).

There is no definitive treatment to prevent, delay or slow the disease process of HD at present (McColgan & Tabrizi 2017), and the disease cycle can last 15 or more years. During this time, symptomatic management might target symptoms such as chorea, rigidity, depression, irritability and altered sleep cycles (Phillips et al. 2008). In drug development for both curative and symptomatic treatment there is still some way to go for effective medical treatment. Due to the longevity and lack of cure, long term management of HD is required and there is increasing recognition of the importance of non-pharmacological management. Engaging in meaningful activities may be important in HD where quality of life both in terms of physical and psychosocial well-being, are severely affected (Helder et al. 2002). Supportive resources to engage in physical activity (PA) for example could be important and relevant, especially resources that are robustly underpinned by appropriate theory.

The argument for PA for people with neurodegenerative conditions is supported by research in PD and MS. Positive influences of PA have been observed specifically in PD and MS in terms of activities balance confidence, fatigue, PA levels, functional measures, measures of gait and health related quality of life (Miyai et al. 2002; Caglar et al. 2005; Ashburn et al. 2007; Kargarfard et al. 2012; Schenkman et al. 2012; Learmonth et al. 2013; Guidi et al. 2013; Gervasoni et al. 2014). Indeed, this is relevant to HD because of the shared characteristics with HD. In HD specifically, there exists a growing body of evidence that highlights the potential that PA may have in symptom management, improved quality of life and disease modification. A number of exercise programmes delivered as part of small scale clinical feasibility studies have resulted in improved strength, health related quality of life and achievement of functional goals in HD (Khalil et al. 2012; Busse et al. 2013; Quinn et al. 2014) (appendix 1).

Zinzi et al. (2007) were the first to identify potential benefit of a non-pharmacological intervention involving physiotherapy. Significant effects were seen in motor and functional assessments but design limitations highlighted the need for more robust future investigations. Subsequently, Kloos et al. (2013) conducted the first controlled trial of an exercise programme with a cross-over design. Participation in an interactive video game led to improved dynamic balance during walking in individuals with HD. However, lack randomisation meant that causation by a third factor linked to both intervention and outcome could not be ruled out. There was also lack of time between interventions to avoid carry over effects, and lack of information about whether the positive effects transferred functionally to daily activities. At a similar time, the first randomised controlled exercise study of a community based exercise programme in HD indicated a significant difference in a mental health component of a quality of life questionnaire (SF-36) (Ware & Sherbourne 1992). The intervention comprised aerobic and strengthening components. More recently, another randomised controlled trial of exercise in HD demonstrated the most significant findings to date (Quinn et al. 2016). Intervention group participants had improved fitness, disease specific motor scores, and improved cognitive score following exercise programme completion compared to control participants. This intervention was more intensive than that delivered by Busse et al. (2013) and participants had the choice of completing it at home or in a local gym with tapered one to one supervision (Quinn et al. 2016).

In the studies by Busse et al. (2013), Kloos et al. (2013) and Quinn et al. (2016) there were no social control comparator groups so it is possible that positive social interactions in addition to, or rather than the exercise, influenced these findings. Busse et al. (2017) addressed this issue in their larger scale feasibility study of an exercise

intervention (ENGAGE-HD), by including a social comparator group and exercise intervention group. Increased self-efficacy for exercise and increased levels of PA were identified in participants who completed the PA intervention and not participants in the social comparator group. Considering this, and potential disease modifying benefits based on animal models and literature in other neurodegenerative diseases, an emerging picture of the importance of specific support for PA in HD is becoming clear.

In addition to their findings, Busse et al. (2017) successfully responded to the need for more robust, theoretically underpinned exercise intervention research in HD. Self-determination theory (SDT) was used to develop the PA intervention (Busse et al. 2017). The PA intervention included one to one coaching in a style that promoted autonomy, competence and relatedness (key components of SDT). A key part to the PA intervention was the tailoring of it to meet individual needs, which, given the complexities of HD would be appropriate. A PA workbook which could be personalised was developed in consultation with HD families and caregivers (Busse et al. 2014). It was advantageous to have a framework because theory (SDT) guided the intervention, and therefore how the intervention was developed was transparent. Also, the use of theory to develop the intervention takes into consideration the MRC guidance regarding the need for use of theory in development of complex interventions.

All of the exercise research studies discussed involved people from early to mid-stage HD and delivered different interventions, over different lengths of time. This suggests the possibility that different modes and PA participation may be beneficial, thus giving people with HD more choice about what they could participate in to gain benefit. This could be favourable because of the heterogeneous nature of HD that causes different people to have different needs and abilities which would be important in considering PA participation. In comparison to Zinzi et al. (2007), the studies by Busse and Quinn (Busse et al. 2013; Quinn et al. 2016) included comparator control groups and assessments were blinded, reducing the possibility of assessor bias and retention over the whole study was good. Importantly, despite the limitations of small sample size, what these feasibility studies demonstrate is that it is acceptable and safe for people with HD to exercise in their local gym or at home with the appropriate support.

Through the exercise studies in HD, an evidence base is developing that clinicians, health and care professionals can use to justify their practice in encouraging people with HD to be more active. The importance of self-management, and in particular in HD, has been of increasing focus in England and Wales with the continuing rising prevalence of long term conditions (Murphy et al. 2010; NICE 2014; Wales Audit Office

2014). Following the government White paper of 2012 (Health 2012), 'no decision about me, without me', drivers have been directed towards achieving more and improved shared decision making within the NHS.

Key points within the White paper that outline how shared decision making should be improved include patient and caregiver involvement in decisions about care and treatment, and supporting the patient to take more responsibility for what they can do to maintain and improve their health. As such, an emergence of 'Expert Patient Programmes' (EPPs) has been seen in England and Wales. EPPs are designed to educate and empower patients with the aim of improving quality of life for those living with long term functionally limiting conditions and reduce burden on health services (Wales 2012; Department of Health 2013). No such programme is currently available for HD. The complexities of HD in terms of the psychiatric, psychosocial and cognitive issues in addition to the movement disorder would require consideration, which the generic courses led by volunteers are not designed to do. Indeed, the need for better expertise of healthcare professionals (HCPs) has been identified by people with HD and their caregivers (Skirton et al. 2010; Maxted et al. 2014; Røthing et al. 2014). Providing advice about self-managing specific to PA at a simplistic level would likely not be appropriate for people with HD and better understanding of the impact of HD on PA participation is needed to underpin development of such interventions.

The use of theory to underpin such resources or interventions is crucial; a criticism of previous interventions has been a lack of theory to underpin them (Jones et al. 2009). Busse et al. (2016) argue that using theory to underpin development of their 'ENGAGE-HD' intervention will ultimately facilitate translation into clinical practice. Indeed specific support for self-management approaches (underpinned by theory) in other neurological conditions such as the 'Bridges Programme for stroke' have been successfully developed and implemented in recent years (McKenna et al. 2015). Large costs associated with long term access of healthcare services and of needs being unmet over time have contributed to the conception of such programmes (McKenna et al. 2015). The 'Bridges' programme which is underpinned by social cognition theory (SCT) takes a holistic approach to encouraging self-management. For example, in the Bridges programme setting personal goals which may include PA is incorporated as a core programme component. (Personal goal setting is discussed in SCT (Bandura 1998). As such, it is apparent that setting relevant personal goals are helpful in engaging people with the programme (McKenna et al. 2015). Given the current lack of a cure and limited means of treating HD symptoms, PA may provide people with HD and their families with an alternative additional option to help

manage the disease. The growing evidence base in support of PA and acknowledgment of the need for theoretically underpinned interventions suggests that better understanding of the impact of the nuances of HD on PA participation is needed. Although the subjective experiences of HD and PA need to be explored to gain better knowledge and understanding, much is already known about the progression of physical, cognitive and behavioural symptoms of HD. Considering these symptoms in relation to PA will help to build a clearer picture. Therefore, the following sections consider the stages of HD and how the symptoms may influence how people engage and participate in PA.

### **3.3.3. The stages of Huntington's disease and potential impact on physical activity (PA)**

In this section the stages of HD are discussed in more detail in light of current knowledge of HD. Within each stage the researcher considers the potential impact of HD on PA at each stage given the growing evidence base for potential benefit of regular PA in HD indicated in the previous section 'current management of HD and potential for self-management through physical activity'.

#### *3.3.3. Prodromal-stage Huntington's disease and potential impact on PA participation*

When an individual receives a positive gene test (they have confirmation of the mutated Huntington gene), but no symptoms are clinically observable, they are considered to be 'prodromal'. The term 'pre-manifest' was previously used to describe this stage but suggests that at this stage there is no evidence of disease. Recently, existence of pathological changes in the brain, behavioural symptoms and neurocognitive signs have been found years before a clinical diagnosis (which is based on motor symptoms) (Stout et al. 2011; Tabrizi et al. 2013). A longitudinal study using brain MRI scans (Tabrizi et al. 2013) concluded that although not mirrored functionally, pathological changes due to HD were present in people considered 'pre-manifest'. The investigators suggested that compensatory networks allow individuals to continue functioning normally up to a point. However once too much pathological damage has occurred, a failure of these compensatory networks results in the development of motor symptoms seen at early stage.

Apathy, irritability, perseverative behaviour, depression and difficulty with complex thinking tasks have been described as characteristic of this stage and reported in HD before manifest motor symptoms (Rosenblatt & Leroi 2000; Kirkwood et al. 2001). Irritability resulting in a reduced control over temper which may result in verbal or behavioural outbursts could potentially impact an individual's PA experience. Such

outbursts could make an individual socially undesirable and impact on PA for the individual in a social setting. Apathy is defined as lack of motivation not attributable to a diminished level of consciousness, cognitive impairment, or emotional distress (Marin et al. 1991). It has been argued that apathy is a salient pathological feature of prodromal HD, often mistaken for ‘laziness’ or depression by family members (Klöppel et al. 2010). Apathy can be present in the absence of depression and is defined as a lack of initiative, loss of interest, lack of perseverance to complete tasks and reduced spontaneity (Marin et al. 1991). Dynamics within spousal relationships change even at this stage. Spouses may take on a more supportive role if there are mild cognitive impairments (Decruyenaere et al. 2003). This may also apply to PA, where if the affected person regularly exercises, their spouse may have to support them. Particularly being reminded to do things as specified by Williams et al. (2012) may include reminders to do their activities. The Apathy Evaluation Scale (AES) is a validated and reliable scale which measures apathy by treating it as a psychological dimension defined by “simultaneous deficits in the overt behavioural, cognitive, and emotional concomitants of goal-directed behaviour” (Marin et al. 1991). Studies using this scale have reported different prevalence of apathy in HD, ranging from 14% to 84-99% (Naarding et al. 2009; Reedeker et al. 2011; Thompson et al. 2012). This could be important as increased passivity as a result of apathy could impact engagement in PA and goal-directed behaviour from early on.

Depression has been identified within prodromal HD and major depressive disorders are seen significantly more than in the general population (Duijn et al. 2008). Julien et al. (2007) argued that the rate of depression increases as a function of proximity to clinical onset. However, following assessment of depressive symptoms in a sample of 803 people with HD Epping et al. (2013) argue that gene positive individuals are at increased risk to develop depressive symptoms at any time during the prodromal stage. Additionally, significant positive predictors of depression include illness perceptions of identity and perceiving the cause to be related to chance; being unlucky to have inherited the gene (Arran et al. 2014). Despite the possible pathologic nature of depression in HD, in light of these findings interventions which target changing illness perceptions could be further explored. A longitudinal investigation of functional decline in HD found that more rapid functional decline was associated with depressive symptomology. The study also found that better neuropsychological and cognitive status at baseline was associated with less rapid functional decline (Marder et al. 2000). It appears that although an individual may still be physically independent, cognitive impairment can be functionally debilitating in HD. A number of small scale studies investigating function have conducted gait analysis of people with prodromal

HD using accelerometers. Decreased velocity, impairment of dynamic balance, increased gait variability relative to stride length variability and swing time variability were detected (Rao et al. 2008; Collett et al. 2014). Although these were small sample studies, they revealed significant gait impairments in people at prodromal stage which are not picked up by less sensitive measures in the clinical setting.

In addition to pathological changes, the impact of receiving a gene positive result may have social and emotional implications leading to self-imposed isolation and certain behaviours (Hagberg et al. 2011). Following genetic testing, participants described trying to do enjoyable things while still healthy which may have implications for PA engagement (Hagberg et al. 2011). This could be a key point at which PA could become an integrated part of their lifestyle. Indeed, the different possibilities of how individuals at prodromal stage choose to acknowledge HD has relevance to PA as health behaviour. If people identify that they are experiencing somatic symptoms and are motivated to problem solve them, contact with health professionals may result in PA being recommended or referral to a physiotherapist who advises on PA. Or if they previously or currently participate in PA, recognition of functional changes may motivate them to continue to try and maintain their function. Those who disengage and deny that anything is changing in terms of their health may be less likely to change anything about their lifestyle. If that were the case, it would seem unlikely that PA behaviour would be motivated by change in health status for such individuals. As this may be a key point at which PA could have an impact on the long term disease pathway, a better understanding of engagement in PA is needed.

#### *3.3.4. Early-stage Huntington's disease*

Progression from prodromal to manifest or early stage HD is recognised with a formal diagnosis of observable clinical motor signs. The time it takes for motor symptoms to develop which lead to a formal diagnosis varies between individuals. The length of the gene mutation and environmental factors influence age of onset and severity of progression (Walker 2007). Progressive onset of motor symptoms such as altered gait, balance and coordination skills are seen and from early stage onwards and people may find it less easy to perform certain physical tasks and activities. In addition, cognitive and or behavioural difficulties can manifest (if not already present) and become more obvious. Although people with HD are sometimes described as not having awareness about their neurological, cognitive and behavioural symptoms, Langavant et al. (2013) have recently shown that at an early stage, patients retain awareness of memory deficits.

Van Duijn et al. (2008) demonstrated a link between increase in irritability and an increase in motor symptoms in people who were previously at a pre-motor symptomatic stage. Van Duijn et al. (2008) suggest that irritability may be an early and sensitive marker for disease progression. Clinical and socio-demographic independent correlates of irritability include CAG repeat, benzodiazepines, being married/living together (Reedeker et al. 2011). Prevalence of irritability in HD for people being married/living together in the study findings of Reedeker et al. (2011) perhaps suggests that irritability may be more pronounced in intimate relationships and may cause more triggers of increased irritability. This is worth considering in the context of PA behaviour and experiences. Although caregivers may be valuable in encouraging and motivating people with HD, the relationship is a complex one and therefore the caregiver may or may not emotionally always be best placed to prompt or encourage PA. Kloppel et al. (2010) and Naarding et al. (2009) both found that depression can be present in early stage HD (in up to 40% of people) (Naarding et al. 2009). They highlight that with disease progression, personality changes become more evident due to dementia and cognitive decline. In a longitudinal study investigating functional decline in HD, more rapid functional decline was associated with depressive symptomology (Marder et al. 2000). Better neuropsychological and cognitive status at baseline was associated with less rapid functional decline (Marder et al. 2000). Anti-depressants used to treat depression in HD can cause side effects including nausea and irritability and dizziness among others. Any side effects of medication may affect a person's ability to participate in PA.

Behavioural dysfunction can be disabling for people with HD and could impede ability to utilise motor or cognitive skills that may still be available in the early disease stages (Hamilton et al. 2003). Although their study was retrospective and of small sample size, Hamilton et al. (2003) make an important link by highlighting the value in assessing behavioural change and awareness which could impact on functional decline.

Disinhibition described in the early stage by Hamilton et al. (2003) may be socially disabling as it could be perceived as anti-social behaviour by others. If for example an individual with HD is participating in PA in public and exhibits what is seen as socially unacceptable behaviour, the person with HD may be asked to leave. In this way, disinhibition may impact on PA engagement.

Compared to healthy control subjects, cognitive impairment has been demonstrated in people with HD. These include deficits in executive function (planning, sequencing, prioritising, organisation, adapting), memory, verbal fluency, psychomotor speed, reasoning, perceptual problems, and delay in acquisition of new motor skills (Bäckman

et al. 1997; Walker 2007; Dumas et al. 2013). Such cognitive impairment early on could affect engagement in PA that requires adapting to new routines, performing new complex tasks, and planning and prioritising chores or activities throughout the day so that time is given to incorporate PA. Using new equipment or being pragmatic about how to use equipment may be difficult (Marder et al. 2000). Those with memory problems may not remember to attend exercise classes. In particular, recall of instructions for using equipment may be difficult for people with HD, but recognition is easier (Pollard 2008). Watching others carrying out the task may trigger the memory of how to do it themselves but this is still to be explored. If people at early stage recognise that they have problems with their memory this may mean that they are amenable to receiving prompts and reminders about daily activities such as PA (de Langavant et al. 2013).

If instructions are being given or the given activity relies on verbal communication, the person supporting the person with HD to exercise would need awareness of the difficulties people with HD may have in responding to questions or instructions immediately. Difficulty in following instructions may come across as indifference causing the people they are working with to lose patience or interest if the instructor is unaware of HD symptoms. This is reflected in the findings by Hartelius et al. (2010) who found that people with HD perceived a need for conversation partners who could adjust and understand potential communication difficulties. It may be challenging for the person with HD to communicate their ideas or needs and therefore communication may need to be adapted in performing certain activities. Hartelius et al. (2010) found that feeling safe and supported, adjustment by the conversation partner, focused questions, taking part in activities and old memories were perceived to have positive effects on communication and to be stimulating.

Perceptual problems in HD include impairments in emotional recognition, perception of time, smell identification, spatial perception and unawareness. There may be an inability to recognise one's own disabilities and therefore exhibit unsafe behaviours (Nance et al. 2011). This may have implications for people being realistic and safe about what PA they participate in. Furthermore, visuo-spatial skills deficit can result in bumping into objects while walking which can cause falls and injuries (Rosenblatt et al. 1999) and would also need to be considered during PA. Impairment of complex types of attention make dual-tasking in HD difficult and ability to complete daily living tasks such as dressing, shopping and managing finances can be greatly affected (Lemiere et al. 2004).

Chorea describes sudden irregular involuntary movements in the extremities, over time progressing to facial grimacing, trunk shoulder and neck movements (Nance et al. 2011). Chorea is prominent in the early and mid-stages of HD, can peak at 10 years post onset, plateau and then lessen, or it may worsen as patients enter late stage. With progression, bradykinesia and rigidity can become the more functionally disabling motor symptoms (Nance et al. 2011). Chorea causes increased postural sway during walking where the limits of stability are exceeded thereby causing falls (Grimbergen et al. 2008) and Quinn et al. (2013) found that people with HD adopt a wider base of support in standing. Changes in gait (Rao et al. 2008; Collett et al. 2014) may cause increased trips and stumbles which may make an individual more aware of their condition or warier when carrying out functional tasks including PA. It is also important to note that falls are a risk at any time including during PA (Grimbergen et al. 2008; Busse et al. 2009; Kloos et al. 2010 and Williams et al. 2014).

In a study of 24 people with HD, Busse et al. (2009) found that those reporting less activity were more recurrent fallers. Falls in HD have been linked to reduced PA where people who are less active fall more often (Busse et al. 2009). However there is discrepancy regarding prevalence and pathophysiology of falls in HD (Grimbergen et al. 2008; Busse et al. 2009; Kloos et al. 2010; Williams et al. 2014). In an observational study, 14 of 24 participants were classified as recurrent fallers (58.3%); 2 or more falls in the previous year (Busse et al. 2009). Kloos et al. (2010) reported a prevalence of less, with 36% being recurrent fallers (more than one fall in previous six months) amongst a cohort of 94 patients with HD. Whereas 60% of patients were classified as recurrent fallers (reported two or more falls in the preceding year) by Grimbergen et al. (2008). Despite the small sample sizes and short lengths of time for prospective data for these studies, the difference in findings may reflect how HD manifests differently between individuals (Walker 2007; Ross et al. 2014). Nevertheless, it is important to ensure safety measures and risk assessments are in place to deal with falls risks such as obstacles on the floor (Grimbergen et al. 2008) so that people with HD are not prevented from participating in PA. This is especially important considering that most falls rarely result in major injuries (Grimbergen et al. 2008).

Peavy et al. (2010) recorded falls and cognitive ability in early HD and found an association between cognitive decline and function. Falls were recorded retrospectively (12 months) and prospectively (3 months). Cognitive scores were significantly lower in fallers compared to non-fallers (Grimbergen et al. 2008). Of the falls reported, one third occurred while multiple tasks were being performed simultaneously suggesting that multi-tasking causes additional challenges. Indeed, Fritz et al. (2016) found that people

with early to mid-stage HD have impairments in cognitive-motor dual-task ability related to functional ability.

A small feasibility study found that people with HD are also more sedentary; they had a reduced daily step count compared to population norms (Busse et al. 2013). Stepwatch activity monitors were used to measure daily step counts and percentage of time a person was sedentary or engaged in PA (Busse et al. 2013). Stepwatch activity monitors are vulnerable to user error in that they have to be worn correctly to collect accurate data which may place limitations on the findings. Busse et al. (2009) suggest further investigation is required to explore whether reported reduction in PA is a result of the motor impairments in HD or perceived balance and confidence. Considering the plethora of symptoms associated with HD it is apparent that a reduction of PA could be equally due to behavioural consequences such as apathy, or cognitive impairment. It is important then to consider that people with HD may experience barriers to keeping active, associated with cognitive and behavioural impairments; which may explain sedentary behaviour (Elsworth et al. 2009; Quinn et al. 2013). In developing the research presented here, disease specific barriers are a consideration; the nuances of how HD symptoms may influence PA participation are yet to be explored.

### *3.3.5. Mid-stage Huntington's disease and potential impact on PA participation*

At this stage, a worsening of HD symptoms is often marked, leading to decreased independence and reliance on others for activities of daily living. This stage is also identified as a critical period for contemplating suicide (Paulsen et al. 2005). Movement disorders characteristic of HD which manifest clearly by mid-stage can include rigidity and motor impersistence chorea and dystonia and or akinesia, hypokinesia and bradykinesia (Mahant et al. 2003; Walker 2007; Goldberg et al. 2010; Quinn et al. 2013). These symptoms may make it difficult for people with HD to co-ordinate and perform specific movement patterns or sustain a particular posture in performing PA. Motor symptoms lead to gait impairments and postural instability which could impact on an individual's ability to engage safely in activities.

A cross sectional observational study found that decreased stepping response time was associated with lower balance confidence scores (Goldberg et al. 2010). Other observations indicated bradykinesia in the form of slowed gait, prolonged reaction time, slowed movement of the upper extremity, reduction in speed of first step of ambulation and longer reaction time in HD participants. These impairments could impact various aspects of PA. Ability to participate in sports where speed and agility, ability to change direction and maintain balance and quick reactions are challenged could be

compromised. Gait impairments include altered gait pattern comprising shorter and more variable stride length and slower speed. Despite findings that falls are common in HD and people who fall have lower balance confidence scores, the fear of falling does not appear to be a common feature (Grimbergen et al. 2008). This could be due to lack of falls leading to serious injury. It might also suggest that people with HD decline from PA participation for reasons other than fear of falling. Loss of independent mobility and functional decline occurring as a consequence of balance and gait disturbances and motor impairments have been reported as critical predictors for placement in nursing homes (Wheelock et al. 2003; Quinn et al. 2013). Wheelock and colleagues assessed motor function (including chorea, bradykinesia, gait abnormality, and imbalance) of people with HD who were living at home and in 'skilled nursing facilities'. Those who were living at the nursing facility had worse motor function and it was found that these motor variables alone predicted institutionalisation.

Dystonia, is a repetitive, abnormal pattern of muscle contraction associated with a twisting quality and often emerges in mid to late stage HD. Dystonia results in asymmetric postures and movements, including arm elevation when walking, trunk tilting and foot adduction when walking (Nance et al. 2011). Rigidity usually manifests at later stages of HD and causes an increase in muscle tone and reduction of passive range of movement. As a result of these progressive impairments people with HD experience clumsiness and awkwardness in carrying out manual tasks. A significant reduction in lower limb muscle strength has been reported which would impact on ability to use certain equipment (Busse et al. 2008). A hand held dynamometer was used to assess isometric muscle strength of six lower limb muscle groups in twenty people with HD and healthy matched controls (Busse et al. 2008). On average those with HD had approximately half the strength of healthy matched controls. Precision grip control which is important for manipulation of objects has been shown to be altered and highly variable in people with manifest HD (Rao et al. 2011). Altered precision grip control (Rao et al. 2011) may significantly impact ability to use particular equipment during PA. Inability to perform activities requiring such control may have a negative effect on the confidence of somebody with HD, which may lead to avoidance of the activity.

Motor impersistence describes the inability to maintain a voluntary muscle contraction at a constant level (Walker 2007) this can cause dropping of items, difficulty with writing and manual tasks, and can prevent effective use of a walking aid. Ability to perform physical activities could be affected by manifestation of motor impersistence and rigidity causing clumsiness and awkwardness in carrying out tasks. Ability to perform

any PA requiring fine motor skills could be affected. If we are aware of these experiences (as healthcare professionals or even family members/caregivers), strategies to address these issues may help maintain successful participation, however there is currently a gap in this understanding of what people with HD actually experience.

### *3.3.6 Late-stage Huntington's disease*

Motor symptom progression in the advanced stages of HD severely limit mobility and ability to carry out activities of daily living. Most people will require full assistance, relying on nursing care. Severely limited mobility and falls limit people functionally. In addition, postural changes, pain, skin breakdown, and respiratory limitations can occur at late-stage (Busse et al. 2008). Chorea and dystonia may become more prominent but usually manifestation of rigidity and bradykinesia (Nance et al. 2011) result in more Parkinsonian like movements where the individual is slow to initiate and terminate movements. Movements are generally slower and stiff due to rigidity of limbs and trunk at this stage. These symptoms would make it difficult to engage in forms of PA that are not adapted to the individual's functional abilities. Particularly later on, the impact of chorea on mobility is such that assistive devices including wheelchairs are used for the individual's safety because of falls risk (Carlozzi & Tulsky 2013).

Global dementia develops with progression of cognitive symptoms. Psychiatric deterioration continues but it is thought that some comprehension is retained (Rosenblatt & Leroi 2000). Impaired speech makes communication increasingly difficult, which could have implications for PA participation, possibly in terms of safety. At this stage, most out of home placements occur. Wheelock et al. (2003) found that people with HD institutionalised in nursing homes have more severe psychiatric and behavioural symptoms than those living at home, and more impaired motor function. At this stage, PA as it is traditionally thought of may not be possible, but active assisted movements and functional activities to preserve quality of life may be helpful.

There appears to be paucity of research conducted with people with late stage HD. With recognition of changes in the brain even at prodromal stage there is emphasis of research on modifying disease progression at earlier stages. A combination of this and the fact that HD is considered rare disease may explain the current lack of research in late stage HD.

### *3.3.7. End-stage Huntington's disease and potential impact on PA participation*

People at end stage HD are likely to be living in a nursing home with mostly palliative care being given or at home with full nursing care as they become increasingly

incapacitated. People become unable to coordinate movement and lack strength to manoeuvre a wheelchair and so become completely mobility dependent on others. At this stage people are usually confined to a nursing bed because of the inability to maintain a safe sitting posture due to rigidity, lack of body control and postural control (Quinn et al. 2012). Twenty-four-hour assistance is required with all aspects of daily living due to loss of meaningful functional movements and ability to wash, dress and eat independently (Stanley et al. 2015).

When difficulty eating becomes problematic causing aspiration and weight loss, a Percutaneous Endoscopically-placed Gastrostomy tube (PEG), or Nasogastric Tube may be fitted so that patients can receive the calories and nutrients needed (Stanley et al. 2015). Bladder and bowel control is lost and ability to communicate is impaired by the associated dementia and motor symptoms affecting speech and restricting ability to interact, understand and perceive. Speech and meaningful functional movement and cognition become almost or completely impaired. Death follows from complications such as choking, infection, or heart failure, typically 15 to 20 years after onset of HD (Walker 2007).

As with late-stage HD, there is paucity of research into end-stage HD generally. Access to people at this stage of HD may be difficult, but also capacity to consent and actually participating could be an issue due to the cognitive deterioration which may be why there is a lack of research. Typical palliative care approaches may include physiotherapists working with the multidisciplinary team. At this stage of disease, any activity would need to be completely supported. Examples of treatment aims include reducing pain and promoting comfort, reducing risk of skin breakdown and infection, minimising risk of aspiration and maximising any retained movements. Passive movements, active movements, active assisted movements, positioning and passive joint range of motion may be considered a type of activity which are used to work towards treatment aims (Quinn et al. 2012).

This section ‘HD: the biomedical perspective’ has enabled the researcher to elucidate the physical, behavioural and cognitive consequences of HD across the stages. More literature exists regarding prodromal and early to mid-stages. This may be due to logistical and ethical issues that would be associated with carrying out research with people at end-stage because of the issue of capacity to consent and severe physical and cognitive limitations. In brief, considering the potential benefits of regular PA participation and the plethora of issues that could potentially influence engagement in PA, a better understanding of how people with HD experience and perceive PA is

needed. The following sections focus on existing literature regarding experiences of living with HD and experiences of PA in HD.

### **3.4. Huntington's disease from the personal perspective**

This section presents literature regarding experiences of living with Huntington's disease (HD). The previous section focussed on the pathological effects of HD on the body and their medical and functional consequences. This section focusses on how those consequences affect daily living from the perspectives of the people living with HD, which includes patients and their familial caregivers. To gain a holistic picture of how HD impacts daily life, it is important to gain an understanding of personal challenges, perspectives and experiences that have been identified in current literature. Such understanding of the literature will help identify issues that may be relevant to PA participation which could be further explored through this research i.e. identify gaps in knowledge which may be relevant in terms of PA participation. In addition, a better understanding of the personal experience of HD may inform research design in terms of considerations and logistics of data collection, sampling, and identifying topics that may contribute to the research questions.

Following the 'PEO' approach, database search terms included (P/E) 'Huntington's disease', 'Huntington's chorea'; (O) 'experiences', 'perceptions', 'views', 'qualitative'. Initially, limits placed on the searches included full text, English language, and papers published within the last 10 years. As this search turned up very few papers the publication date limit was altered to include papers from the year 2000 onwards. A small number of papers were identified which were screened further by title and abstract. This resulted in a number of papers within the literature focussing on experiences of people with HD that pertained to illness perceptions (Helder et al. 2002a; Kaptein et al. 2006; Arran et al. 2014), communication (Hartelius et al. 2010), health related quality of life (Carlozzi & Tulsky 2013), experiences of healthcare (Skirton et al. 2010; Etchegary 2011) and issues surrounding genetic testing for people at risk of HD (Hagberg et al. 2011; Hagen 2017) (Hagberg et al. 2011). Other literature pertaining to living with HD focusses on the 'HD family'; support provided by familial caregivers and how roles within the relationship change over time with progression of HD (Helder et al. 2002b; Kaptein et al. 2007; Downing et al. 2010; Aubeeluck et al. 2012; Williams et al. 2007; Maxted et al. 2014.; Røthing et al. 2014).

Some research studies used qualitative methods such as interviews and focus groups. Most studies used questionnaires and surveys to obtain perceptions and experiences.

Responses are limited to answering specific questions and participants are guided through the questions in a very structured way. Another potential limitation of the literature is that most of the research has been conducted outside the UK. Although it is not the nature of qualitative research to be generalizable, specific cultural differences may mean that the same research carried out within a UK context may provide different results. There generally seems to have been limited research carried out about experiences of living with HD in a purely qualitative sense. However, the existing literature does provide some helpful insight into the impact of HD on those living with it.

### **3.4.1. Characteristics and challenges of Huntington's disease**

A significant dilemma that people in HD families face is whether to have the genetic test to confirm whether they have inherited the mutated HD gene. Decruyenaere et al. (2003) longitudinally investigated psychological distress in the 5-year period after genetic testing in HD. The authors suggest that self-protective mechanisms such as denial or avoidance may be a sound way of keeping stress and anxiety at manageable levels, and escape from pessimism. Changes in relationship dynamics and changes in roles and responsibilities in the home were identified following thematic analysis. For example, how a spouse would talk to their husband or wife to avoid confrontation. Also giving spouses reminders to do things they had forgotten, and 'swapping' of the main income earner due to the person with prodromal HD being unable to continue full time employment. It is clear that different individuals have different ways of dealing with the situation. These findings suggest that spouses can take on a more supportive role even at prodromal stage (Decruyenaere et al. 2003). Hagberg et al. (2011) used semi-structured individual interviews to explore the physical, social and emotional impact for participants who had taken a genetic test 5-14 years' prior and were gene positive. Most participants described relief from uncertainty about knowing whether they carried the gene or not. Additionally, perspectives of new uncertainties following the positive result such as uncertainty about onset of symptoms and how the disease would manifest, but also trying to do enjoyable things while still healthy were highlighted. Such uncertainties relate to what Hagen (2017) described as the fluid and dynamic experience of HD following their study that focused on the genetic aspect of it. Semi-structured interviews were carried out with family members not at risk of carrying the HD gene (n=5), people who tested negative (n=2), one person who was at risk, and people who tested positive (n=3). Interpretative phenomenological analysis revealed that noticing symptoms in everyday life and participants comparing themselves to other family members contribute to the complexity of trying to create a stable lived experience of HD (Hagen 2017).

It is particularly interesting in this study that Hagen (2017) picked up on the way that participants interpreted everyday happenings within a framework of HD. Prior to gene testing, participants felt the presence of HD in everyday life, whenever they stumbled when walking or their fingers twitched, even though two of them subsequently had negative test results. In light of their findings, Hagen goes onto discuss the importance of case by case healthcare that considers the individual's experiences of HD. What would have been additionally interesting to explore is if a follow up with participants who received a positive result revealed an influence on health related behaviour, which may have implications for PA participation.

A study using an illness perceptions questionnaire found that people with HD characterise it as an illness of long duration, with lack of cure or possibility of symptoms improving (Helder et al. 2002a). Furthermore, in a study that explored the perspectives of people across the stages of HD regarding personal concerns, the emotional, social and identity related concerns were similar over the course of the disease (Ho & Hocaoglu 2011). Concerns about physical issues appeared to manifest at early stage (mirroring onset of motor symptoms which confirm a diagnosis) and became more dominant over the course of the disease. The questionnaire led interviews were limited to only identifying the concerns and did not extend to trying to further understand the reasons for the concerns. Arran et al. (2014) suggest that people with HD have a strong illness identity and perceive themselves to have little control in a personal sense and in terms of treatment. These findings echo the current biomedical knowledge of HD in terms of a lack of cure and limited medical interventions to control symptoms. However, despite the fact that there is no control over stopping HD, people with HD may exercise control over their lives in other ways. For example, Arran et al. (2014) suggest that having small goals as a way of making life more meaningful may empower people to regain perceived control.

Beliefs about the illness appear to play a role in well-being and so enhancing an individual's perception of control on some level may be important to their quality of life. Having a sense of control may be elicited through coping strategies, which, in addition to illness perceptions have been linked to well-being in HD (Kaptein et al. 2006). Behavioural disengagement, mental disengagement, focus on and venting emotions, and strong illness identity were negatively related to patient's well-being (Helder et al. 2002a). Alternatively, coping strategies related to acceptance of HD have been positively related to well-being (Helder et al. 2002a).

Variation in coping mechanisms have been reported (Helder et al. 2002b; Arran et al. 2014). Arran et al. (2014) found that participants reported more use of emotional and instrumental support and less planning and positive reframing (finding the positives in situations) in comparison to those in the study by Helder et al. (2002a). The two studies used the same questionnaire (Illness Perceptions Questionnaire) but a different version. One version accounts for specific context of the individual (used by Arran et al. 2014) whereas the other is more generic, so could account for the different emphasis on use of various coping mechanisms.

Carlozzi and Tulsky (2013) found that people with HD in their focus groups reported feelings of loneliness, hopelessness, emotional avoidance (denial) and guilt about passing on the gene to their children. They also demonstrated resilience in terms of appreciating life, having hope and wanting to keep going for as long as they could (Carlozzi & Tulsky 2013). The way in which people react to living with HD could depend on a number of reasons, from their previous experiences to how the HD is affecting them, and their beliefs. Chorea was perceived to be more distressing for others to see rather than being distressing and disabling to the persons with chorea (Carlozzi & Tulsky 2013). Carers highlighted this by describing how wheelchairs were used for people with altered gait due to chorea because of safety concerns regarding falls. Motor functioning and activities of daily living were of concern to people with HD, the caregivers and clinicians. Activities of daily living not only rely on physical functional ability; experiences of reduced ability to communicate can cause difficulties in everyday life and lead to social isolation (Saldert et al. 2010).

Focus groups with people with HD and family members highlighted experiences of difficulty with language comprehension, speed of communication, depth of conversation, and increased effort and concentration required during communication in a study by Hartelius et al. ((2010). This is reflective of the findings by Saldert et al. (2010) that people with HD experience difficulties such as initiating conversation and understanding of complex discourse, which could lead to reduced interactions with others and social isolation. These issues caused by cognitive decline could clearly have implications for the experience of PA. The experience of communication being challenging for people with HD is also something to consider when collecting data which relies on them taking part in discussions. As Hartelius et al. (2010) included caregivers in their focus groups this approach could also be used to explore the impact of HD on PA. Hartelius et al (2010) allowed the caregivers to respond on behalf of the care recipient. Although this would be the caregiver's interpretation rather than that of the person with HD, they can provide insight into and recognise additional issues that

people with HD do not have, because of the cognitive decline associated with the disease process.

### **3.4.2. Experiences of healthcare services**

Skirton et al. (2010) investigated experiences of healthcare services from the family caregiver point of view because they are generally most involved in all aspects of care, often while bringing up children and working. Responses from a postal questionnaire identified carer concerns about health care services for their family member. Family member caregivers perceived that healthcare professionals lacked understanding of HD and individualised care was highlighted as an important concern as well as lack of supportive community resources.

In another study which aimed to explore healthcare experiences of HD families, Etchegary (2011) conducted semi-structured interviews with members of families affected by HD. Similar to Skirton et al. (2010), lack of knowledge of healthcare professionals and difficulty gaining access to appropriate care services were highlighted as concerns. Understanding of the complex and evolving needs of the individual with HD and their family members (who may be their caregivers) appears to be an important need that is not met satisfactorily. Improved confidence in healthcare professionals' abilities to help could influence help seeking behaviour and alleviate some of the family member caregiver burden. Although there is paucity of research in this area, the evidence indicates that improved knowledge and understanding of HD is needed and how HD families might be supported. Rarity of HD could possibly be attributed to lack of healthcare professional experience of medical management of HD but improving their knowledge would help. Etchegary (2011) suggests that better education for healthcare professionals is needed regarding the complex nature of HD.

### **3.4.3. The 'HD family'**

Family members reported changes in the general functioning and social relationships of individuals with HD even from prodromal stage in a focus group study (Williams et al. 2007). Family members of people at prodromal stage highlighted experiences of emotional outbursts. Secrecy surrounding the positive gene test meant that outbursts could not be explained to others. Also, the worry of how being gene positive for HD would affect legal rights was a driver for maintaining secrecy. Themes identified describe the changing roles of the family members having to care for the person with HD and how they prepare for and manage that. The changing dynamic between spouses and how people try to maintain 'spousal relationships' highlight the struggle and tension of trying to retain what was once normal. Family members described how their spouses or family member with prodromal-HD would 'put on a role' in public that

was different to when they were at home with regards to anger, mood and irritability. Hence alterations in cognition, behaviour, and functioning observed by family members may not be obvious to others at this stage (Williams et al. 2007). This may have implications for qualitative exploration of PA experiences if there is conflict between what the family member and person with prodromal HD perceive, or if the family member feels restricted in disclosing their thoughts for risk of upsetting the person with prodromal HD. Interestingly in another study, partners attributed changes in the person with HD to other factors (Downing et al. 2010).

Downing et al. (2010) found that people with prodromal HD and their partners attributed functional changes to aging and other health conditions, work environment, and temperament. Avoidance of making HD related attributions may be interpreted as denial. Denial is a recognised coping mechanism in HD where there is no cure and treatment is limited (Decruyenaere et al. 2003). Since avoidance and disengagement may impact on help seeking behaviour this may negatively influence well-being over time. Downing et al. (2010) suggest that denial may be detrimental, also highlighted by Helder et al. (2002a) who stress that problem focused coping (such as seeking medical help) is paramount considering that symptoms will undeniably develop over time. It would be helpful to know whether healthy lifestyle behaviours such as PA have any relevance for people coping with HD or the knowledge that they will develop HD at this early stage. Across the literature, people with HD attempting to maintain the feeling of control relating to the impact of the disease was identified. Gaining knowledge about HD to make informed decisions and exercise choice, and familiarising themselves with ongoing research which may give hope for the future have been elicited as strategies for maintaining control. Other strategies identified include asking for support and using humour to deflect from difficult situations (Maxted et al. 2014).

In a study with 51 couples (people with HD and their spouses), Kaptein et al. (2007) used the Illness Perceptions Questionnaire (IPQ) to explore ‘illness perceptions’ of the participants as described by Levanthal et al. (1984). The IPQ consists of subscales of “Identity”, “Timeline”, “Cause”, “Consequences”, and “Cure/Control”. Kaptein et al. (2007) argue that illness perceptions and influence of caregivers could have implications for quality of life of the person with HD. This is because illness perceptions are related to how people react and cope, and spouses become increasingly responsible over time for the daily activities of the person with HD. Perceptions of illness duration, curability and causality did not differ significantly between people with HD and their spouses. However, people with HD were found to hold more positive beliefs than their partners regarding symptoms on the ‘identity’ subscale of the

questionnaire and more perceived control than their partners. Kaptein et al. (2007) suggest further research to elicit what the actual contributions of spouses' perceptions are in terms of outcome for the patient. Nevertheless, their findings perhaps indicate the importance of having the spouses on board with management or treatment options including PA.

Maxted et al. (2014) found that HD impacts on family dynamics such that identities change and roles within the family shift. Phenomenological analysis of interviews with family dyads (seven parent/adult child dyads) revealed a 'protective' and 'supportive' dynamic where families safeguarded themselves as a unit. This 'unit' could serve as a barrier to others including healthcare professionals and other family members outside of the 'unit' but also lead to feelings of entrapment for those within it. HD is so multidimensional and complex because of the symptoms, psychosocial implications, who is affected and how (beyond the individual being treated) and how the effects on others within the family impacts back on the individual being treated. Caregiver burden in HD has previously been identified to have a cost to caregiver health and quality of life (Williams et al. 2009; Aubeeluck et al. 2012; Cox 2012). Family members have expressed their difficulties in coping with a disease that is always evolving (Aubeeluck et al. 2012) and have spoken about grieving for loss of a previous life and adapting to a new one (Williams et al. 2009). Lack of time to themselves has been found to be a recurring issue for caregivers of people with HD (Williams et al. 2009; Aubeeluck et al. 2012; Cox 2012). In addition, caregivers report being resigned to not receiving the professional support required (Aubeeluck et al. 2012). Knock on effects of these issues could be detrimental to the care, well-being and health of the individual being cared for. Maxted et al. (2014) and Røthing et al. (2014) suggest that when working with people with HD, healthcare professionals should be cognisant of the HD family context and take a 'family system perspective'. Changing the family dynamic through 'individualistic' therapy could lead to isolation of the person with HD. In managing an individual with HD, considering the wider context of the individual to incorporate family members would be more conducive to a holistic treatment or management approach.

Literature about the personal perspective of HD provides insight into potential personal issues, summarised in the following section, which may impact on their ability to participate in PA but also to participate in research. Design considerations for this research may relate to caregiver burden, people's motivations for participating in the research and managing expectations. Such factors will need consideration when developing a plan for data collection location, how to sample, the information received by participants, and the researcher's role in consenting and explaining the research.

#### **3.4.4. Summary of ‘Huntington’s disease from the personal perspective’**

The existing literature pertaining to HD portrays a formidable, devastating disease that overshadows the lives of the individuals who have it and the families who live with it. Lack of a cure means that current treatment consists of managing symptoms and therapeutic counselling. It appears even accessing appropriate healthcare is an ongoing challenge for people with HD and their family caregivers (Skirton et al. 2010; Etchegary 2011; Aubeeluck et al. 2012; Maxted et al. 2014; Røthing et al. 2014). Physical activity (PA) has been shown through a growing evidence base to have potential positive influence on symptoms and psychological issues associated with HD. PA is something tangible and people may take control of for themselves and may temper the feeling of entrapment and loss of control described by people with HD (Arran et al. 2014; Maxted et al. 2014). However, the multidimensional and complex nature of HD could make PA engagement challenging despite the potential benefits that could be gained, especially as sedentary behaviour has been highlighted early on (Busse et al. 2013). The importance of using theory to underpin further exploration and develop understanding to develop supportive resources for PA in HD has been highlighted as an important component (Jones et al. 2009; Green 2014).

The evolving needs of the individual with HD which impact on them and their caregivers as people progress through the stages could have implications for engaging in PA. The heterogeneous nature of HD means that not every individual experiences all of the symptoms described, but a combination of symptoms could cause complexities in activity participation. Further understanding how people living with HD experience PA could be important in helping them to engage in PA and overcoming barriers to avoid sedentary behaviour. In the following section the current literature exploring experiences of PA in HD is reviewed.

### **3.5. Current literature exploring experiences of physical activity in Huntington’s disease and other long term neurodegenerative conditions**

#### **3.5.1. Overview**

The purpose of the following section is to highlight current knowledge of physical activity (PA) experiences in HD and to identify gaps in that knowledge that the researcher may address and contribute to moving this knowledge forward through their research. Following a literature search described below, only four qualitative studies of

PA in HD were identified. Due to the lack of literature and its limitations in regarding exercise experiences in HD, the search was broadened to include other long term neurodegenerative conditions. The literature reviewed therefore included research pertaining to experiences of PA in Parkinson's disease (PD), Multiple Sclerosis (MS) dementia and Alzheimer's disease (AD).

The above mentioned conditions result in similar symptoms or have similar consequences to HD in terms of body structure and function, activity and participation. For example, all are progressive conditions as with HD. PD and MS are both progressive neurological diseases in which movement disorders manifest and result in reduced mobility and function. Behavioural difficulties and cognitive impairment is seen in AD and dementia which negatively affects physical functioning and abilities to carry out tasks of daily living. Reduced independence and PA are seen in PD, MS, dementia and AD (Jankovic 2008; Chertkow et al. 2013; NICE 2014). Extending the literature review to include these conditions allowed for issues around PA to be highlighted that could be relevant for people with HD. The terms 'Parkinson's disease', 'Alzheimer's disease', 'dementia' and 'Multiple Sclerosis' were added as key search terms to the original database search. This resulted in a further fifteen studies being identified for the literature review (appendix 5).

A PEO approach was used to create search terms to identify relevant literature. Where 'P' represents patient, population, problem; 'E' is the exposure or issue; 'C' is comparison (if any) and 'O' is outcome (Bettany-Saltikov 2012). Key search terms used were (P) 'Huntington's disease', 'Huntington's chorea'; (I) 'exercise', 'physical activity'; (O) 'experiences', 'perceptions', 'views' and 'qualitative'. The Critical Appraisal Skills Programme (CASP) approach was used to appraise the identified literature (CASP 2014).

Database searches were carried out using ASSIA, Pubmed and Allied and Complementary Medicine, Embase 2000 to 2016 February 29, Ovid MEDLINE(R) without Revisions 1996 to February Week 4 2016, PsycINFO 2000 to February Week 4 2014, PsycArticles Full Text, Pubmed and Applied Social Sciences Index and Abstracts (ASSIA). The following inclusion criteria were used to identify relevant literature: 'full text', 'English language' and date limit of the year '2000' to current. Further refinement of the literature involved reading the abstracts of those papers identified from the searches to see whether they were relevant. At this point papers were excluded if they were irrelevant (did not explore experiences of PA or exercise in HD). Hand searching

of reference of the relevant papers to identify further research papers was also carried out.

The four studies identified from the literature search for qualitative research related to exercise experiences in HD report common barriers to different exercise experiences and contribute to a small evidence base (Zinzi et al. 2009; Quinn et al. 2010; Khalil et al. 2012; Frich et al. 2014).

### **3.5.2. Experiences of physical activity in Huntington's disease**

Three studies related to experiences of specific exercise intervention programmes in HD (Zinzi et al. 2009; Khalil et al. 2012; Frich et al. 2014). Quinn et al. (2010) conducted interviews with people with Parkinson's disease and early to mid-stage Huntington's disease who had participated in independent exercise programmes. The other study that explored experiences of PA in relation to a structured exercise intervention is the randomised controlled trial that the researcher supported in their role as research assistant (Busse et al. 2013). The process evaluation findings have been published in an abstract (Debono et al. 2012) and the researcher has access to the full unpublished data. The qualitative aspect of the study was limited in what it captured in view of what the researcher had witnessed in delivering the exercise programme, (described in the introduction chapter). Despite this, the findings provide insight in addition to the limited literature that exists and so the findings are reviewed here also. Most recently, another study conducted focus groups with people with HD to find out what would be helpful from a PA workbook that they were developing for participants of a randomised controlled trial (Quinn et al. 2016). Please see appendix 4 for more details of the papers reviewed and appendix 3 for the CASP table.

The studies identified highlight perceived physical and psychological benefits of participating in the exercise interventions. In particular, these were reported as perceptions of improved balance, feeling fitter, weight loss, increased energy levels, improved ability to perform exercises at the gym sessions or to complete walking sessions, improved gait, increased confidence and self-esteem and feeling of relaxation and improved mood (Debono et al. 2012). It would perhaps be interesting to further explore what these perceived benefits mean for people's everyday lives as there was little discussion of this by the authors. Common barriers described by the studies include lack of motivation and physical difficulties due to the disease. (Zinzi et al. 2009; Quinn et al. 2010; Khalil et al. 2012; Debono et al. 2012; Frich et al. 2014). Whilst some studies elicited disease specific issues (cognitive and physical) impacting on PA participation (Zinzi et al. (2009; Khalil et al. 2012), not all participants of the

study by Quinn et al. (2010) identified disease specific issues as key barriers. This perhaps highlights different insights that people have into their condition and different presentation of symptoms. Lack of insight is not a salient feature of HD but can be present (Quinn et al. 2010). The barriers highlighted appear consistent with the possibility of apathy presenting at early stages of HD and manifestation of motor symptoms from early to mid-stage HD. Quinn et al. (2010) found that most participants had never been provided with exercise-specific information relevant to their condition which was perceived as another barrier to PA participation.

Support and individualised PA were perceived facilitators to participation (Zinzi et al. 2009; Quinn et al. 2010a; Khalil et al. 2012; Debono et al. 2012; Frich et al. 2014; Quinn et al. 2016). Issues caused by HD (elicited in the section 'Huntington's disease from the biomedical and personal perspective') such as difficulty learning and remembering new tasks, remembering appointments, social isolation and preferring routine appear to tie in with the concept that having one to one support and prompts were helpful for participants to adhere to the programmes. Participants of the community based exercise intervention used diaries as prompts, reminders and monitoring tools (Debono et al. 2012). Support from family members bringing participants to the gym sessions was vital to their adherence. Family members provided moral support and encouragement. They also gave helpful reminders and positive feedback about participant's achievements if they noticed changes with functional activities such as walking.

Khalil et al. (2012) found that self-efficacy was influenced by participants' belief in their ability to use the DVD and that they could influence their health. The link between self-efficacy and adherence to PA has also been documented in other neurodegenerative conditions (Ellis et al. 2011; Elsworth et al. 2011; Kasser & Kosma 2012; Eriksson et al. 2013). The support of an instructor in one of the interventions in particular was perceived to improve confidence and ability to go to the gym which was a new and intimidating environment initially (Debono et al. 2012). Further consideration of self-efficacy in the context of HD and PA may help understanding of how HD impacts on PA and could be important considering the helplessness that people with HD often described (Helder et al. 2002a).

Quinn et al. (2010) and Frich et al. (2014) specifically highlight the concept of participating with others with HD. Participants of the inpatient rehabilitation programme reported social benefits of being part of an 'HD group' (Frich et al. 2014). In contrast, interview participants in the study by Quinn et al. (2010) expressed concern at seeing

other people with the same disease particularly if they were at more advanced stages. The differences observed are perhaps reflective of the importance of personal preference for people with HD engaging in PA. Various reasons could affect preferences for example; self-perception; how people are coping with the HD, previous experiences or personal beliefs and motivations to exercise (Quinn et al. 2010) but this needs further exploration.

In considering motivation, a number of the studies found this to be positively influenced by an understanding of the purpose of exercises, goal setting and collaboration in developing a personal exercise programme rather than being given a prescribed routine (Quinn et al. 2010; Frich et al. 2014). In comparison, Debono et al. (2012) found that motivation was facilitated through family support and the presence of an instructor. The studies exploring PA experiences in HD are generally well described with clear aims and transparent, rigorous methodologies. The pragmatic approach of using qualitative research to evaluate specific exercise interventions for HD is of course very helpful to help improve future development and design of specific interventions. Findings mainly discuss barriers, facilitators, perceived benefits and the issue of motivation. However, exploration of the participant experiences relates to specific programmes; home-based exercise, inpatient rehabilitation. That is, the research is limited within the context of that specific intervention with people who fit the inclusion criteria for that interventional study. It is therefore unlikely that broader issues outside of such structured programmes associated with PA participation in HD would have been captured. Furthermore, the breadth of experiences being explored would be limited because participants had chosen to take part in the PA interventions. Nevertheless, although the studies investigated different types of intervention, recurring themes in relation to barriers for participant engagement are seen and begin to form a thread of what may be pertinent to PA in HD (Zinzi et al. 2009; Khalil et al. 2012). Indeed, an important facilitator to adherence appears to have been support.

Khalil et al. (2012) identified strategies used by caregivers such as prompting and cues that helped participants to overcome barriers, highlighting the need to understand more about the role of caregiver input for PA. However, the studies were not designed to consider in depth experiential exploration of meanings, perceptions and nuances of daily living with HD and how that effects their PA experiences. A good example of this is the interview schedule for the COMMET-HD trial, which the researcher was involved in. The schedule was designed to specifically explore participants' experiences of different aspects of the exercise programme including gym sessions, independent walking sessions and interactions with the supervisor in the gym sessions. Questions

asked about barriers and facilitators that promoted or challenged adherence to the intervention, social support and strategies that helped participants to adhere to the programme. This is common across the studies; they focussed on evaluation of the interventions that participants had completed and generally explored PA experiences in HD.

The studies by Zinzi et al. (2009), Debono et al. (2012), Khalil et al. (2012) and Frich et al. (2014) have a clear process for reporting adherence and good adherence to the exercise programmes was achieved. This is interesting as evidence suggests that people with HD would experience more barriers to PA participation and that they are more sedentary compared to population norms (Busse et al. 2013). Indeed, the process evaluation identified numerous barriers including the social environment of the gym, impaired gait, falls, tiredness, other commitments including holidays, caring for other family members, hospital appointments and domestic chores (Debono et al. 2012). The observable movement disorder which people may be embarrassed about and the fact that people with HD can be mistaken for being 'inebriated', may help explain concerns about the social environment (Maxted et al. 2014). Capturing data that would help explain why adherence was good, could have been informative to future intervention design. However, the studies' designs were limited in being able to explore the underlying mechanisms of impact, interaction between participants and the interventions or the influences of context on intervention implementation (Moore et al. 2015).

Qualitative methods facilitate investigation of how participants experience interventions and understanding of the context in which they are delivered; key to understanding why interventions succeed or fail (Anderson 2008). Although the studies indicate that people with HD believed that exercise improved well-being and might keep them stronger, functional and fit, few in the study by Quinn et al. (2010) participated in regular exercise. Knowledge of the people living with the disease could offer valuable insight into living with HD and how it impacts on PA interventions as has been seen in other conditions such as Parkinson's disease (PD) and Multiple Sclerosis (MS) (Jones et al. 2008; Elsworth et al. 2009; Dlugonski et al. 2012) The interventions were successful in engaging people in the particular contexts of delivery. Whether such interventions would lend themselves to be replicated successfully elsewhere and how external factors might influence delivery and function is unknown. This is because the exploration was focussed on the specific context and intervention. For example, support within the structure of the exercise programmes in terms of cues, simple instructions, diaries, individualised support from a knowledgeable instructor, physical

support, social support and social interaction leading to psychosocial benefits were perceived to facilitate participation, motivation and improved well-being (Zinzi et al. 2009; Quinn et al. 2010; Khalil et al. 2012; Debono et al. 2012; Frich et al. 2014). In trying to understand context, qualitative approaches may be particularly useful because of the ability they provide the researcher with to explore social or behavioural processes where quantitative methods are limited (Lewin et al. 2009).

The MRC framework 2008 (Anderson 2008) emphasises the importance of qualitative research to achieve better developed interventions and better designed evaluations, i.e. interventions that could be successfully implemented in clinical practice and quality research (evaluations) facilitated by effective design. The process evaluation guidance recognises that the interaction between participants and the intervention will be shaped by participant attitudes, beliefs and pre-existing circumstances. Contextual factors can act as moderators of intervention outcomes; the same intervention in a different context may have different outcomes. The process evaluation guidance published recently suggests that quantitative and qualitative analyses can “build upon one another”; where qualitative data can be useful in explaining quantitative findings and quantitative data may be used to test hypotheses developed or generated by qualitative data (Moore et al. 2015). The use of theory and qualitative research at numerous stages during the research process is suggested (Moore et al. 2015). Perhaps reflective of a lack of theoretical guidance, the themes from the studies reviewed here come across relatively generic in line with the general exercise literature in healthy populations, for example ‘facilitators and barriers’.

Use of theoretical frameworks to underpin the studies reviewed could have provided them with a focus through which to guide the research as described by Green (2014). Only one study attempted to use theory to understand their findings. Quinn et al. (2016) interpreted their findings broadly within the framework of self-determination theory which discusses motivation as existing along a continuum from intrinsic motivation (for personal fulfilment) to extrinsic motivation (not having personal control) (Teixeira et al. 2012). Paucity of theoretical frameworks used to underpin design or analysis of the findings may explain the leniency of the study findings towards rather generic themes. Using a theoretical framework may have helped unpick experiences of HD and PA more meaningfully specific to HD. For example, from the earlier review of HD literature there is reason to suggest that people with HD may not experience PA in the same way as those in the healthy population and therefore, themes that reflect the specific nature of HD may be more useful in understanding their experiences of PA. Previous research exploring experiences of HD has been embedded in theoretical frameworks (Kaptein et

al 2007; Arran et al. 2014; Helder et al. 2002) (Appendix 6) that have been found to be relevant in furthering understanding of HD, but not been explored in the context of PA. This exposes a gap in the current evidence base for PA in HD.

In spite of their limitations, the studies are the first to draw attention to the experiential nature of PA in HD and so the findings are of significance in terms of adding to the knowledge base. There are no studies that consider PA in prodromal or end stage HD. This may be because the evidence base is still being developed or difficulty of accessing people at prodromal stage who may not want to engage with the idea of HD, and the ethics associated with including participants at end stage who are severely cognitively impaired. The findings from existing studies seem to suggest that although the programmes were perceived to be acceptable by most participants, there were mixed experiences. For people with HD, engaging in PA may be complicated by the triad of motor, cognitive and behavioural symptoms that are experienced over time.

Prior to symptom manifestation, people with HD may well engage in exercise and then continue to exercise despite disease onset. However, they may need to develop strategies to deal with how the symptoms affect their abilities (cognitively, behaviourally or physically). Clearly evidenced by the lack of research studies identified, this has not been explored in HD. As subjective experience has such a bearing on whether people engage in PA, it is important that these subjective experiences are explored. Further exploration to understand the features of HD and how they impact on PA participation and experiences is needed to give healthcare professionals the best chance to encourage PA in HD for the benefits that could be gained. Due to limited literature in HD it is helpful to consider experiences of exercise in other long term neurodegenerative conditions that have similar symptoms. This is considered in the next section of this chapter.

### **3.5.3. Experiences of physical activity in other neurodegenerative conditions**

#### **Overview**

Due to the lack of experiential research regarding HD and PA the purpose of this section is 1) to present background knowledge of experiences of PA in other conditions that have relevance to HD in terms of the symptoms and their impact and 2) highlight the use of theoretical models underpinning the research so as to inform this research.

A PEO approach was used to create search terms to identify relevant literature. Where 'P' represents patient, population, problem; 'E' is the exposure or issue; 'C' is comparison (if any) and 'O' is outcome (Bettany-Saltikov 2012). Key search terms used were (P) 'long term neurological condition', 'neurodegenerative condition' 'Multiple

Sclerosis', 'Parkinson's disease' and 'Alzheimer's disease'; (E) 'physical activity', 'exercise'; (O) 'experiences', 'perceptions', 'views', 'exploration' and 'qualitative'. The Critical Appraisal Skills Programme (CASP) approach was used to appraise the identified literature (CASP, 2014).

Database searches of ASSIA, Pubmed and Allied and Complementary Medicine, Embase 2000 to 2014 February 29, Ovid MEDLINE(R) without Revisions 2000 to February Week 4 2014, PsycINFO 2000 to February Week 4 2014, PsycArticles Full Text, Pubmed and Applied Social Sciences Index and Abstracts (ASSIA) were carried. The following inclusion criteria were used to identify relevant literature: 'full text', 'English language' and date limit of the year '2000' to current. Further refinement of the literature involved reading the abstracts of those papers identified from the searches to see whether they were relevant. At this point papers were excluded if they were irrelevant (did not explore experiences of PA or exercise in MS, PD or AD). Hand searching of references of the relevant papers to identify further research papers was also carried out.

From the literature search, 15 relevant studies were identified in Multiple Sclerosis (MS), Parkinson's disease (PD) and Alzheimer's disease (AD). Of the 15, seven were studies exploring experiences of specific exercise programmes that participants completed as part of larger exercise trials (Dodd et al. 2006; O'Brien et al. 2008; Smith et al. 2009; Plow et al. 2009; Eriksson et al. 2013; Learmonth et al. 2013; Skår et al. 2014). Of those, two studies explicitly used theoretical theories/models including the transactional model of stress and coping model and social cognitive theory (Plow et al. 2009; Eriksson et al. 2013). The other eight studies were stand-alone qualitative studies aiming to further understanding of the experiences and meanings of PA for people with MS, PD and AD (Jones et al. 2008; Elsworth et al. 2009; Ravenek & Schneider 2009; Kayes et al. 2011; Smith et al. 2011; Dlugonski et al. 2012; Cedervall et al. 2015; Malthouse & Fox 2014). Of those, two used the World Health Organisation International Classification of Disability and Function (WHO-ICF) (Jones et al. 2008; Ravenek & Schneider 2009) and one used social cognitive theory (Kayes et al. 2011).

In the qualitative studies that were linked to quantitative studies of an exercise intervention, authors were generally explicit about their involvement in intervention delivery and conducting the interviews. Most describe a thorough and transparent process of data collection, analysis, and evidence of reflexivity threaded throughout the study design. For example, in one study, consideration is given to who conducted the interviews, i.e. not the same person as who delivered the exercise programme (O'Brien

et al. 2008). Themes across the literature relating to physical activity (PA) experiences in other long term neurodegenerative conditions can be broadly attributed to the overarching concepts of barriers and facilitators to participation. Disease specific barriers were identified as influencing PA in terms of symptoms, stigma, and external barriers (Jones et al. 2008; Elsworth et al. 2009; Smith et al. 2011; Dlugonski et al. 2012; Skår et al. 2014; Cedervall et al. 2015). Across the studies, facilitators to exercise in PD, MS, and AD were described as different types of support, enjoyment, perceptions of taking control for health through PA, the social aspect of exercising with similar others, self-efficacy and overcoming barriers. Swimming, stretching, walking and group exercise sessions were perceived to be enjoyable and beneficial. Perceptions of symptom improvement were also motivating for participation in exercise (Smith et al. 2009; Smith et al. 2011).

Given the possible cognitive and physical symptoms associated with MS, PD and AD, achieving engagement in PA is a more complex undertaking for people with these conditions, however many positive outcomes of PA participation were also reported. Positive outcomes included psychological, physical and social factors such as strength, walking, and feelings of normality, friendships and participation (Skår et al. 2014). Participants of a resistance strengthening programme with MS reported being in the group environment with others with MS gave them more confidence and feeling of being safe in trying new things (Dodd et al. 2006).

In particular, the importance of social support to participate in PA from family, friends, healthcare professionals and others with the same condition is highlighted in PD, MS and AD. In a study exploring social support influences of PA participation participants felt more supported by physiotherapists, exercise trainers and massage therapists than neurologists and general practitioners should give more encouragement for PA (Ravenek et al. 2009). This indicates the potential for physiotherapists to positively influence PA participation, which may be relevant for HD too. A limitation of the study by Ravenek et al. (2009) is that all participants interviewed were currently active. The experiences and factors influencing participation in exercise may be different for those with PD who are inactive. Nevertheless, in light of this being the first study to explore factors influencing PA participation in PD, the data could be informative for strategies to increase PA participation in HD. Certainly, there are advantages to including participants who have experience of regular exercise for the insight they can give into maintaining participation despite disease specific barriers. For example, Smith et al. (2009) reported that participants spoke about 'staying away from the edge' in the same way that participants describe 'not crossing the line' in the study by Kayes et al. (2011)

in knowing the limit of what they should do and how it may impact on MS related fatigue.

Informational, emotional and physical support were described as important for PA for people with neurological conditions by Elsworth et al. (2009) who conducted four condition specific focus groups with people with MS, muscular dystrophy, motor neurone disease and PD. Again, informational support (defined as advice or knowledge given by other people) was a positive influence on exercise behaviour in PD by Ravenek et al. (2009). 'Emotional support' (encouragement or consoling actions of others) and 'instrumental support' (actions by others that directly facilitated participation) were also found to be key to PA participation (Ravenek et al. 2009).

Considering that caregivers can play an important role in supporting PA, it is important to recognise the issues that they may also face. Malthouse and Fox (2014) found that people with AD did not often link their decreasing cognitive abilities to limits on participation, but the caregivers recognised the increased demands on them with progression of their spouse's condition (Malthouse and Fox 2014). The suggestion by Malthouse and Fox (2014) that the focus should be on encouraging and maintaining active lifestyles instead of structured exercise considers the need to account for the progressive nature of AD. Perhaps having a less stringent approach to exercise may relieve burden caregiver burden and make it more enjoyable to support PA for their family member. Indeed, participants were more likely to continue with leisurely activities that they had always enjoyed, especially if it was something both of the couple had enjoyed.

In exploring the personal experiences of everyday walking in PD, Jones et al. (2008) excluded participants who had taken part in the PA intervention but had cognitive difficulties. Although there is an argument that cognitive impairment may cause inconsistent recounting of events, excluding people who may have communication difficulties further marginalises an individual who may be disempowered due to impairments caused by their condition and have a limited voice in everyday life. Also the research would not be capturing the perspectives of each individual equally based on the communication difficulties perceived by the researchers, which there may well be strategies to help with but which may require some flexibility on the interviewer's behalf (Lloyd et al. 2006). Caregiver's knowledge and understanding of the individual with PD was key to supporting their PA (Jones et al. 2008). They used strategies that they knew worked for the individual and consequently, walking was an enjoyable and relaxing experience for those with PD, which facilitated their continued engagement in

outdoor walking. Two key strategies for addressing the challenges when walking were monitoring walking using concentration and correcting walking. Caregivers were found to help promote participant confidence, and provide encouragement and relaxation particularly in outdoor walking situations. They were able to do this using strategies developed from their knowledge and experience of the participants' problems. The use of the International Classification of Functioning, Disability and Health as a guiding framework helped Jones et al. (2008) to explore the physical, social and environmental aspects of PA in PD.

Aspects of self-esteem such as independence and confidence have also been reported to influence exercise behaviour in other studies in PD (Ellis et al. 2011; Eriksson et al. 2013). Knowledge of the people leading exercise sessions was highly valued by participants with MS (Dodd et al. 2006). Unfortunately, this is not the case for all. In this regard Elsworth et al. (2009) report participant experiences of embarrassment and lack of confidence in fitness professionals due to lack of knowledge about their condition and suitable, safe exercises. Due to the rarity of HD, and lack of knowledge of healthcare professionals already identified in the literature review, this may well also pose a risk for people wanting to do PA in HD but is yet to be explored, highlighting a gap in current knowledge.

Studies have also revealed negative experiences of receiving support from others such as frustration due to lack of empathy or understanding of limitations for the individual. It appears that support offered by others can sometimes be well intended but misjudged, and therefore perceived negatively. For example, people with MS explained that it could be frustrating for them if people in their social network had a poor understanding of the limits of their physical abilities for PA (Smith et al. 2011). Inappropriate jokes about people's movements when exercising with them were perceived negatively (Smith et al. 2011). Skår et al. (2014) found that people with MS also experience undesired attention due to the visible physical symptoms, feelings of shame and being stereotyped as well as people using strategies to try and conceal the MS amongst 'normals' when exercising.

Noticeably, people who were exercising with others with the same condition described the value of emotional support received from them. This finding occurred most frequently in relation to disease specific exercise programmes (Eriksson et al. 2013; Skår et al. 2014; Learmonth et al. 2013; Dodd et al. 2006; O'Brien et al. 2008). Participants with MS found group exercise to be a positive experience in terms of meeting and exercising with others with MS and elicited feelings of being accepted and

feeling ‘normal’ (Learmonth et al. 2013). This was also reported by Dodd et al. (2006), Smith et al. (2011) and Skår et al. 2014). Learmonth et al. (2013) also found that participants learned from each other about balancing exercise in relation to managing energy levels and fatigue. Participants were happy to attend support groups of people at a similar stage and found them positive and supportive, discussing and encouraging different types of exercise. In a qualitative study by O’Brien et al (2008), 12 people with PD with mild to moderate disability were interviewed using a semi-structured interview schedule developed within a phenomenological framework about an eight-week disease specific exercise programme they had participated in. Despite being an exercise programme designed to improve strength and function, the social interaction and well-being experienced through participating in the exercise programme emerged as being more important to participants. It emerged that people measure success differently; a consideration when developing and tailoring exercise programmes to individuals. Pertinent to the progressive nature of the disease, participants who attended support groups were discouraged from attending if other people present were at later stages of PD; “you see what you have to look forward to becoming”. This is something the researcher will need to consider carefully when designing the methods for this research including how, when and where to collect data, as people with HD may feel the same.

Self- efficacy has been associated with exercise behaviour in the studies reviewed (Eriksson et al. 2013; Ravenek et al. 2009; Ellis et al. 2011). In particular, a study that considered factors associated with exercise in PD found that those with greater levels of self-efficacy were more likely to engage in exercise (Ellis et al. 2011). Another study found that women with MS who exercised regularly had very strong personal meanings and motives for exercising, citing accomplishment as one of the reasons for engaging in exercise (Dlugonski et al. 2012). Accomplishment of activities that are meaningful may have a positive effect on self-efficacy which is something Eriksson et al. (2013) explored. They relate their findings to Bandura’s social cognitive theory and specifically focus on the self-efficacy and modelling aspect and self-regulatory efficacy. The social environment is suggested by Eriksson et al. (2013) to be an important factor in facilitating exercise participation. The research was based on a cohort of people who had gone through a PD specific group exercise programme. Bandura (1997) suggests that self-efficacy is mediated through seeing people similar to oneself being successful in the given activity (in this case exercise) and the greater the ‘assumed’ similarity the stronger the personal beliefs of self-efficacy. The disease specific exercise programmes that people took part in for this research were set up for people with PD so although it gives meaningful insight into exercise experiences the context is specific

and perhaps the social dynamics would be different if the researchers had interviewed people who exercise at their local gym for example open to all members of the public.

A study which used individual interviews to explore and generate understanding of the meaning of physical exercise in the lives of people with PD gained insights which could have implications for strategies to promote exercise. Findings from the studies suggest that participation for the inherent satisfaction that can be gained, and increased self-efficacy can facilitate engagement in exercise in long term neurodegenerative conditions. Smith et al. (2009) found that self-efficacy was important for participants to be able to achieve the 'healthy tiredness' they associated with exercise. Perceived control was associated with the confidence that individuals could 'listen to their bodies' and exercise to a point where they would experience healthy tiredness rather than unhealthy tiredness. Healthy tiredness was characterised by participants as a sense of achievement, better sleep and relaxation, improved strength, stamina and being more productive. Unhealthy tiredness was associated with 'shaky' legs, reduced balance and negative feelings of failure, fear for safety and anxiety. Disease specific issues were considered as barriers to PA for many participants of the studies of MS, AD and PD.

Specific symptoms of each condition that needed to be considered to exercise successfully and safely were highlighted by Elsworth et al. (2009). For those with PD, loss of balance and difficulty in moving around were identified as barriers of concern. Disease specific barriers to PA in AD were described as dementia related barriers including loss of initiative to do activities, impaired body function, emotional and environmental barriers, impaired orientation ability and decreased energy levels (Cedervall et al. 2015). Jones et al. (2008) report that walking was described as requiring intense concentration and was therefore more effortful for people with PD. In this regard, the physical limitations of the disease and also 'thinking' about how to deal with the physical symptoms while performing the action of walking (monitoring) may impact on the experience of PA (Jones et al. 2008).

Other barriers to PA emerged as inaccessibility of exercise facilities, cost, transport to the facilities and limited services (Elsworth et al. 2009). Considering that people with HD may not be working or driving because of the symptoms of the disease, it is possible that they too may experience such barriers to PA. Again, this is yet to be explored across the stages of HD but gaining knowledge regarding this may help identify where support from healthcare professionals may be needed.

Physically active women with MS used strategies for overcoming disease specific barriers, and used PA as a way of asserting control over an unpredictable disease

(Dlugonski et al. 2012). Exercise was considered a positive way of self-managing their condition for people with MS to gain long term health benefits and achieve goals. People chose PA that helped them to feel normal, i.e. they were able to do with success. In consideration of these findings, perceived control might facilitate longer term participation if people feel they are able to control their illness through exercise. In addition, perceptions of control are a facilitator to PA in MS (Smith et al. 2009). Indeed, Smith et al. (2011) focus on the concept of self-integrity whereby individuals maintain a sense of self-worth and perceived health particularly through continuing to fulfil social roles despite an illness.

In AD, for some, being physically active was a way of maintaining a sense of continuity through doing something they used to do before diagnosis (Cedervall et al. 2015). Overcoming barriers to continue PA also gave participants a sense of capability. Practical, mental and avoidance strategies to overcome the barriers were described by participants (Cedervall et al. 2015). In conditions where cognitive impairment is known to manifest, attempts to maintain attributes and keep up social roles associated with health and striving to maintain certain roles may be misinterpreted as an ‘unawareness’ of difficulties or limitations. This could be interesting to explore in HD regarding how people may try to continue doing a certain PA as they did before HD, or prior to further progression of HD, and whether they adapt or appraise how successful they are in doing so and the implications of that.

In their study Plow et al. (2009) identified examples of self-regulation such as problem solving behaviour and symptom related barriers. Different people had different perceptions of how active they were and different perceptions of what being active is. Some active participants with MS felt they had control over the disease but some felt helpless as well as in control at times (Plow et al. 2009; Smith et al. 2009). Interestingly and something which may have implications for HD, less active people had more negative coping styles, and were more stressed. The concept of a ‘tipping point’ was elicited in relation to people who are active and inactive where the barriers are perceived too big to overcome to engage in PA.

The following section summarises findings from reviewing the literature relating to experiences of PA in HD and other neurodegenerative conditions and how this has guided the researcher’s thinking about their research.

### **3.5.4. Summary: current knowledge of experiences of physical activity in Huntington's disease and other neurodegenerative diseases**

The studies reviewed explore the experiences of various types of exercise in different environments and highlight the impact of those differences. For example, Khalil et al. (2012) found that people with HD exercising at home using an exercise DVD experienced lack of motivation as a barrier to doing exercise. Barriers perceived during a walking programme for people with PD included being self-conscious in public, busy unpredictable outdoor environments and required concentration on others and concentration on one's own actions causing tiredness and being a frightening experience for some (Jones et al. 2008). Khalil et al. (2012) and Jones et al. (2008) found factors to be prominent in individual or caregiver assisted exercise that were different in comparison to group exercise experiences in PD, (Eriksson et al. 2013) highlighting the psychological difference between exercising in a group situation to individual or caregiver assisted exercise.

Prominent factors in group exercise included positive impact on various aspects of quality of life related to 'social life', 'meeting others similar' and experiencing 'fellowship' categorised as strengthening one's self- image linked to coping strategies and being part of activities linked to having explicit life goals. Where lack of motivation and being self-conscious were identified as salient issues by Jones et al. (2008) and Khalil et al. (2012), these did not emerge in the group exercise setting for Eriksson and colleagues (2013). Perhaps it was experiencing positive social interactions with others with PD that helped to overcome any self-conscious feelings by seeing similar others and participants were motivated by the group setting as has been seen in MS (Skår et al. 2014). It may be more difficult to be motivated or feel empowered to do the activity if there is nobody to encourage or share the experience. It is clear from the researcher's review of the literature that in neurodegenerative conditions, different PA will have different psychological effects and as with PD, the physical symptoms of HD may impact on experiences in different environments, whether indoor, outdoor, and alone or in a group, in a structured supervised programme or independent exercise. As demonstrated in the literature in other long term neurodegenerative conditions, (Jones et al. 2008; Smith et al. 2009; Smith et al. 2011; Dlugonski et al. 2012; Skår et al. 2014) different strategies can help depending on the PA situation. Strategies to facilitate participation included monitoring and correcting walking, monitoring fatigue, concealing the disease to counteract social stigma (by exercising on days when symptoms were not so visible), prioritising and scheduling PA, managing disease-specific barriers, and building social support networks, but these remain to be explored in depth in HD.

Given the lack of literature in this area in relation to HD, further exploration of strategies that support self-efficacy and motivation is warranted considering the potential benefits of engaging in exercise for people with HD, where poor motivation and apathy have been reported as common behavioural features, (Naarding et al. 2009; Krishnamoorthy and Craufurd 2011). In addition, there is a lack of stand-alone qualitative literature whose main purpose it is to explore and describe PA experiences and how HD impacts on PA. With one exception (Quinn et al. 2016), the research that currently exists does not attempt to embed the research in any kind of theoretical framework. The following section will explore application of theoretical models to PA experiences in other long term neurodegenerative conditions and HD in general as there is paucity within the literature about PA in HD.

### **3.6. Application of theoretical models to explore experiences of Huntington's disease and other long term neurodegenerative conditions.**

The final literature review aim was to identify a relevant theoretical model that would be relevant to the context of HD and PA participation. Use of a theoretical model can guide understanding of the phenomena and gives transparency to the perspective from which the researcher is looking at the phenomena. The identified literature focussing on experiences of PA in neurodegenerative conditions more generally provides a range of instances where theoretical models are used to guide and focus the research. The models include the self-regulation model (SRM), social cognitive theory (SCT) (Elsworth et al. 2009; Eriksson et al. 2013), the World Health Organisation International Classification of Functioning, Disability and Health (WHO-ICF), (Jones et al. 2008; Ravenek & Schneider 2009) and the Transactional Model of Stress and Coping (TMSC), (Plow et al. 2009). Within the literature that focussed on experiences and perceptions of HD, the self-regulation model (SRM) described by Levanthal et al. (1984) was used to underpin five studies (Helder et al. 2002a; Helder et al. 2002b; Kaptein et al. 2006; Kaptein et al. 2007; Arran et al. 2014). (See appendix 6 for descriptive details of the studies.) The use of the different models is discussed in the following section. There is only one example where theory has been applied in research exploring PA in HD where Quinn et al. (2016b) used self-determination theory (SDT).

Quinn et al (2016b) describe development and delivery of the ENGAGE-HD intervention to promote PA in HD. They conducted focus groups (FG) style workshops

to establish what would be helpful for people with HD to promote their ongoing participating in a PA intervention. They also asked for feedback on a physical activity (PA) workbook that they had proposed as a supportive component of the PA intervention. Quinn et al. (2016b) thematically analysed the FG data, describing three main themes of 'personal beliefs and motives', 'enablers' and 'challenges'. They related their findings to SDT which has been commonly used in exercise literature to understand exercise behaviour (Texeira et al. 2012). Although Quinn et al. (2016b) articulate that a range of types of motivation were elicited from the FGs, they describe their findings as "broadly consistent" (p73) with SDT and that their findings "may be at least partially explained using this theory" (p74) but do not give an in depth explanation of this. It seems that the authors identified that people with HD have various motivations for exercising and these can be mapped to the different types of motivation described by SDT. From their literature review they identified that motivation is important for adherence and that adherence to PA is a problem in neurodegenerative conditions but there was little further analytical discussion underpinning the choice of SDT in relation to ENGAGE-HD.

The satisfaction of basic psychological needs of relatedness, competence and autonomy influences whether an individual engages in and maintains a particular behaviour according to SDT (Ryan and Deci 2008). The factors that influence satisfaction of these needs are identified by SDT as psychological, social, cultural and environmental. Autonomous behaviour is described as engagement in an activity through one's own volition. Competence is a need that has to be met for internalisation of a behaviour. It is described as having the confidence and skills to change and to engage with the behaviour with a feeling of success and having the ability or support to overcome control related barriers. SDT suggests that competence is facilitated by autonomy in the sense that once people are engaging in the behaviour of their own volition, they are more inclined to learn and apply new strategies. Relatedness is described by Deci et al. (2008) as the need to feel respected, understood and cared for. SDT argues that autonomy, competence and relatedness are vital to be met in order for achievement of self-determined behaviour that is truly volitional and arises from high quality motivation. Maximising an individual's experience of basic psychological needs is thought to facilitate internalisation of regulation of the behaviour and therefore the behaviour is better maintained (Ryan et al. 2008). The different types of motivation are distinguished by the goals or reasons that result in the behaviour or action (Ryan and Deci 2000).

According to the principles of SDT motivation is not unitary where an individual is either motivated or unmotivated, rather it can be described as existing along of continuum of different types of motivation. At one end is intrinsic motivation followed by integrated, identified, introjected, external regulation and amotivation (no motivation) at the opposite end of the continuum. For each type of motivation there are associated processes. For example, extrinsic motivation in the form of external regulation is associated with salience of reward or punishment and is the most controlling form of extrinsic motivation. It is associated with compliance and reactance rather than autonomous behaviour (Ryan and Deci 2000). Intrinsic motivation is viewed as the most autonomous and high quality form of motivation. It leads to self-determined behaviours which are engaged in because of the inherent interest and enjoyment of doing so and not for separable outcomes or external contingency such as pressure or external reward (Hagger and Chatzisarantis 2008). Extrinsic motivation has been used to explain engagement in the behaviour in the absence of intrinsic motivation. Unlike intrinsic motivation there exist different types of extrinsic motivation. These are non-self-determined or partly self-determined and are defined as external regulation and introjected regulation.

In ENGAGE-HD, once SDT was identified as a theory to help underpin intervention development; components of SDT were targeted to promote motivation of the participants, i.e. training was given to coaches to ensure that their interactions with participants promoted autonomy, relatedness and competence. This was detailed in the logic model developed following the focus groups which was used to illustrate development and implementation of the intervention and subsequent evaluation (appendix 7). The logic model is a graphical representation of how the intervention would work, detailing the various inputs, activities, outputs and expected outcomes. It served to make explicit the various interacting elements of the intervention, which, when evaluating the success of the intervention would allow identification of factors that worked well or otherwise.

Conveying the components of the intervention in such detail could in turn promote translation of clinical research into practice. However, there is no clear mapping process of how SDT was identified as the theory to help understand and explain the FG data and how that subsequently fed into intervention development. The FG findings themselves do not appear to feed into intervention development, despite the authors' argument that speaks of the importance that "any theoretical framework is grounded in and relevant to the particular experiences and needs of the specific population" (Quinn et al. 2016b, p.72). This is illustrated by the researcher in appendix 21. Appendix 23

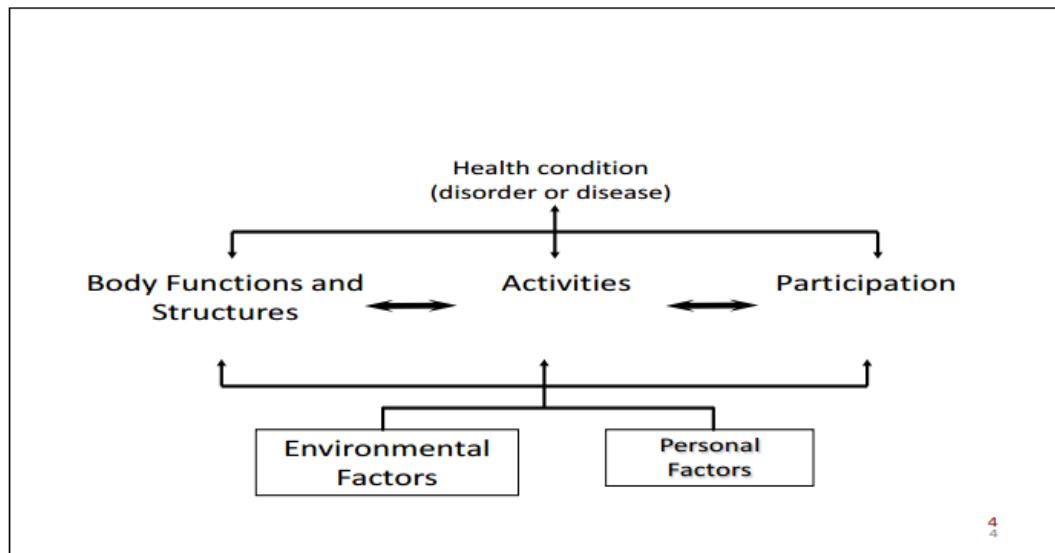
illustrates the process of intervention development that started with the FGs, which were analysed, and themes were identified, but that it is not clear how these findings were integrated into the intervention development underpinned by SDT. Motivation was identified as an issue and it seems that was the driver for using SDT. The figure shows how the idea of different motivations fed into the decision and linked to SDT, which was then used to underpin the intervention consisting of home visits, workbook and DVD that were designed to promote autonomy, relatedness and competence.

The MRC framework for development and evaluation of complex interventions provides an overall rational for a cyclical development process but a criticism of this framework is that there is not sufficient detail regarding each of the steps (Hurley et al. 2016). More detail is given within the process evaluation of complex interventions guidance (Moore et al. 2015), however providing more guidance for development of complex interventions is the six step process created by Bartholomew and colleagues (2016). This process called 'implementation mapping' integrates theory and evidence to design, implement and evaluate complex interventions. As part of the process, behaviours are matched with determinants of behaviour which guides selection of theory based intervention methods. Identification of such determinants other than motivations is not something that appears to have been considered for ENGAGE-HD but could have been, through broader exploration of PA behaviour and experiences during the FGs. Determinants of PA behaviour are something that could have been explored through the FGs, however, from the schedule (appendix 7) it is clear that there were very focused questions specific to developing the intervention, and so there was limited opportunity to do this. (This is similar to the process evaluations of other PA studies in HD that have explored specific experiences of interventions.) It is apparent therefore, that a link which could have further integrated theoretical foundations into the ENGAGE-HD intervention was not utilised. A broader exploration of PA in HD could have resulted in findings which may be applicable to any intervention in terms of determinants of PA behaviour which could then be targeted in interventions. With regards to this research, whilst SDT is useful to explain behaviour specifically from a motivational point of view it does not allow the broader exploration of describing how the nuances of HD can impact on that behaviour and PA participation.

The International Classification of Functioning, Disability and Health (ICF) was developed by the World Health Organisation (WHO) and was endorsed by the World Health Assembly in May 2001 (World Health Organization 2001). The ICF was developed for use in describing functioning and disability in terms of health and health related states and provides a standard language and framework for this purpose (see

figure 3). The ICF can be used in a number of ways and at an individual or population level e.g. as a health classification, as a decision making tool for planning and policy and has been used in research to help understanding of the experience of health and disability (World Health Organization 2001).

Figure 3: WHO-ICF model (World Health Organization 2001)



Functioning is considered from a biological, individual and social perspective by the ICF model of functioning and disability (WHO-ICF 2001). Functioning is conceptualised as dynamic, in such a way that the interactions between health conditions and contextual factors (personal, environmental) are suggested to influence outcomes of disability and functioning. Examples of personal factors considered include age, gender, coping styles, social background, education, profession, past and current experience, and overall behaviour pattern. Environmental factors that comprise the physical, social and attitudinal environment that people live in include support and relationships, attitudes of others, services, systems, policies, products and technology, changes to environment, the natural environment and human made changes to the environment. The outcomes include consideration of 'body function and structure', 'activity' and 'participation'. Anatomy, physiology and psychology of the human body is described by 'body structure and function'. The individual's functional status is described by 'activity' and 'participation', including communication, mobility, interpersonal interactions, self-care, learning, applying knowledge, etc. Both positive and negative aspects of functioning can be explored using the ICF model, therefore providing a multi-perspective approach within the multi-dimensional model.

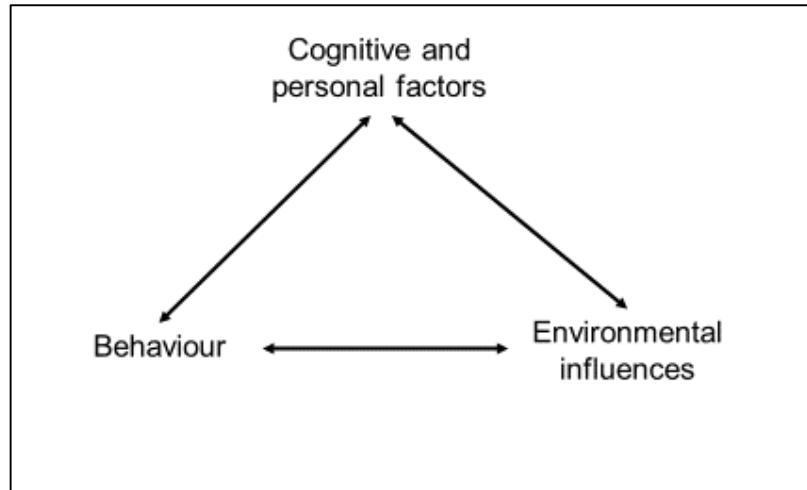
Ravenek and Schneider (2009) and Jones et al. (2008) used the model to underpin their studies of PA in Parkinson's disease (PD). Of particular interest to Ravenek and Schneider (2009) was using the 'environmental factors' component of the WHO-ICF as a 'lens' through which to develop the research exploring PA participation in early stage PD. They achieved this by developing the interview schedule to facilitate exploration of participants' PA participation and its relation to the ICF's stated environmental factors. Social support is considered to be an environmental factor within the ICF. Interview questions developed by Ravenek and Schneider (2009) related to sources of social support considered by the ICF, and sources of social support that had been identified as important in other populations (stroke, MS). These sources included family, friends, and people in the workplace, health care professionals and social support groups. The WHO-ICF was found to be helpful in understanding the experience of PA in the context of early stage PD and environmental influences, with what the authors considered to be a homogeneous sample (Ravenek & Schneider 2009).

Jones et al. (2008) considered components of the WHO-ICF as a guiding framework in their exploration of the personal experiences of everyday walking, challenges and strategies used to compensate for difficulties in PD. Exploring body structure and function, and environmental factors helped identification of challenges experienced with complex walking and key strategies used to address those challenges. The themes were linked to how PD affects body structure and function; i.e. the pathology of PD was linked to the symptoms and challenges shared by participants. The authors concluded that future interventions should allow for the psychosocial as well as physical issues of PD. The study by Jones et al. (2008) links the body structure and function aspects of PD with the functional and environmental factors, and participation in walking, therefore considering the ICF as a whole rather than its component parts, compared to Ravenek and Schneider (2009) who focus on the environmental factors and participation. Although the components of the WHO-ICF facilitate a holistic insight into the human experience of health and disability, when considered as a whole, there is no 'time dimension' to the model. In other words, information gathered regards the individual's health at the particular moment of data collection. However, in a progressive disease such as HD, the ability to capture insight that accounts for the progressive nature of the disease may be more helpful in learning about the condition as a whole and its impact on PA.

Eriksson et al. (2013) (please see study details in appendix 6) relate their findings to social cognitive theory (SCT) (Bandura 1997) to help explain their findings. SCT is named as such because it describes much thought and action as being a result of

social origins and addresses the socio-structural and personal determinants of health. SCT acknowledges that learning is conceptualised as knowledge acquisition through cognitive processing and learning through the effects of one's actions. In other words, humans have capacity to learn vicariously through observing other people's behaviour and the consequences for them, as well as learning through trial and error of their own actions.

Figure 4: Key constructs of Social Cognitive Theory (adapted from Bandura 1986)



Human functioning is described by SCT as comprising key constructs which are: 'behaviour', 'cognitive and other personal factors' and 'environmental events' depicted in figure 4 (Bandura 1986). These constructs reciprocally influence each other in a triadic fashion. According to SCT, interactions of these constructs influence how and why individuals engage in and maintain certain behaviours, of which, personal self-efficacy plays a pivotal role (Bandura 1986).

Personal self-efficacy is described as 'a belief in one's abilities to organise and execute courses of action required to produce given levels of attainments' (Bandura 2000). Bandura describes 'capabilities' of human agency, including forethought capability. This means that individuals can visualise something specific about their future, which leads them to set goals, which then guides their actions in such a way as to achieve that cognised future. For example, in the context of PA, an individual may see themselves progressing the amount of time they can cycle on an exercise bike. They might then set goals such as cycling more often to achieve this 'vision'. An important point made by SCT is that individuals will only act if they believe that they have the ability to achieve the desired outcome or effect through their actions, i.e. without the belief in their ability, they lack the incentive to act. Certain skills may be required to

achieve the desired outcome, and self-efficacy to use the required skills effectively and consistently (even in challenging circumstances) can result in either good or poor use of skills possessed. Strong self-efficacy can be created through mastery experiences (success in something one does), vicarious experiences (seeing people similar to oneself being successful), social persuasion (verbal persuasion that they can do it) and by reducing stress reactions (altering negative emotional proclivities and correcting them). With regard to vicarious modelling, the greater the ‘assumed’ similarity between individuals, the stronger the personal beliefs of self-efficacy. SCT also argues that the more capable an individual thinks they are, the higher the goal they set themselves, the better the strength of commitment to goals and better outcomes they expect for their efforts. Personal self-efficacy can determine perseverance and resilience of individuals when faced with obstacles, adversity and experiences of failure. Self-efficacy can also determine self-hindering or self-aiding thought patterns and the amount of stress and depression people experience in coping with taxing environmental demands. Outcome expectations also play a role in SCT and can be positive or negative, social, physical and self-evaluative. Whereas self-efficacy is the judgement of capability to achieve the desired outcome, an outcome expectation is a judgement of the likely consequence a behaviour will produce; i.e. the outcome is the consequence of a behaviour, not the behaviour itself.

Eriksson et al. (2013) explored the meaning of PA within a social context; with people with PD who had taken part in a group exercise intervention. The social environment was an important factor in facilitating exercise participation. Interviewees explicitly expressed the importance of exercising with and observing similar others for their confidence. Eriksson et al. (2013) relate this to vicarious modelling explained by SCT (Bandura 1986). Confidence in one’s own ability was found to have a positive influence on goal setting and motivation and in turn, on adherence to the programme. This fits with SCT with regard to self-efficacy being mediated through seeing people similar to oneself being successful in the given activity (in this case exercise) and experiencing motivation through meeting goals. Self-regulatory efficacy originates from perceived self-efficacy and is the confidence one has in their ability to influence their behaviour despite difficulties they may encounter (Bandura 1986). Eriksson et al. (2013) discuss the relevance of this in the context of the exercise programme. They found health promoting behaviour to be linked to individual ability to continue with exercise in the face of challenges associated with PD, rather than their mastery of the exercise.

It appears that self-efficacy is relevant to the findings of Eriksson et al. (2013) and others (Elsworth et al. 2009) in understanding exercise behaviours and experiences for

people with neurological conditions other than HD. PD and HD both cause movement disorders, so individuals with either may experience similar difficulties with respect to carrying out exercises and having the confidence to try new exercises or progress. Barriers to exercise for people with PD are also relevant in HD (Elsworth et al. 2009; Plow et al. 2009). If self-efficacy is appropriate in helping understand PA participation for people with PD it could also be relevant for people with HD in engaging in PA. Also in consideration of the wider context of SCT, modelling and increased self-efficacy through seeing similar others succeeding in the specific behaviour / activity may be relevant, however this has not yet been explored in HD and PA.

The researcher particularly wanted to focus on the context of HD, which the WHO-ICF facilitates in terms of the constructs of body structure and function, whereas the SCT provides a strong focus on placing the individual in their social world (as well as their individual world). SCT facilitates exploration of behaviour and motivation to carry out particular behaviours, and it may be interesting and useful to explore the role of the many aspects of self-efficacy in the context of PA and HD, particularly when thinking about intervention development. However, at this early stage of developing the evidence base, a much broader exploration of PA experiences in the context of HD which entails exploration of the social and environmental aspects (and others yet to be identified or explored) may be more appropriate.

In SCT, the focus is perhaps too narrow at this point to facilitate a broader exploration of PA experiences in HD. Another consideration is that people with HD live fairly isolated lives in terms of socialising with other people with HD, especially in the context of PA. Modelling (observing the behaviour of others/vicarious learning) was seen as an important factor to support self-efficacy by Eriksson et al. (2013) but the findings are very context specific. The research was based on a cohort of people who had completed a PD specific group exercise programme. To the researcher's knowledge there are no group exercise classes or PA groups specifically for people with HD in Wales or England which may limit the application of SCT. In common with the WHO-ICF, SCT does not appear to account for change and development over time which may be a limitation in helping to understand the experiences of PA participation over time in HD as the disease progresses.

Plow et al. (2009) used components of Social Cognitive Theory (SCT) and Transactional Model of Stress and Coping (TMSC) in their deductive analysis to explore their utility in understanding PA behaviour among persons with multiple sclerosis (MS). Data was coded into pre-established categories developed using

aspects of SCT and TMSC and explored similarities and differences between people who exercised versus people who did not. The transactional model of stress and coping (TMSC) suggests that interaction occurs between the individual and their environment (Folkman and Lazarus 1984). It describes stress as resulting from an imbalance of demand (pressures) and resources (ability to cope and mediate stress). TMSC was developed as a linear model that describes progression from one stage to the next resulting in emotions and thoughts leading to implementation of a coping strategy. The two stages are described as stages of appraisal. These are: primary appraisal and secondary appraisal of problem based coping (more perceived control over the situation) and emotional based coping (less perceived control over the situation).

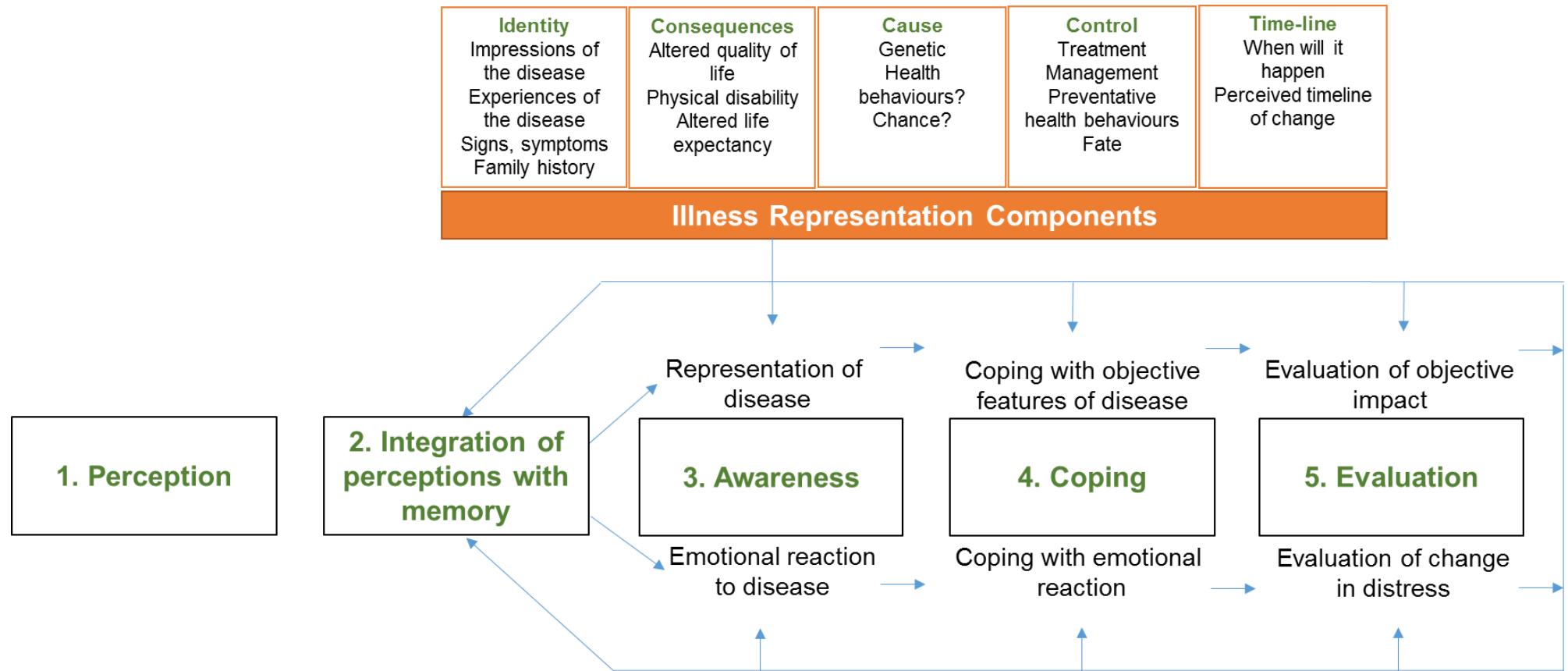
Although constructs of both SCT and TMSC were useful in eliciting understanding of PA behaviour, the fact that the model follows a linear pathway make them less relevant for exploring experiences of PA in HD because of the complexities and multidimensional nature of HD as discussed in sections 2.2 and 2.3 of this chapter. The changes associated with progressive nature of HD means that a more flexible model is required to capture changes that might occur and affect PA over time (Tabrizi et al. 2013). The self-regulation model (SRM) has been previously used to explore experiences of HD. The SRM incorporates the element of time, and was relevant for furthering understanding of the psychosocial aspects and physical consequences of HD (Helder et al. 2002a; Helder et al. 2002b; Kaptein et al. 2006; Kaptein et al. 2007; Arran et al. 2014). The SRM proposed by Levanthal et al. in 1984 was developed through a culmination of preceding research through the 1960s and 1970s that explored how people interpret and cope with health threats (Levanthal et al. 1984). In the database search for literature exploring the experiences of HD, (section 'Huntington's disease from the personal perspective') fifteen studies were identified, and five of those utilised a theoretical model (the SRM) to facilitate understanding of experiences of HD. The SRM was used to explore links between biopsychosocial factors and psychological distress in HD (Arran et al. 2014), illness perceptions, coping and well-being in people with HD (Helder et al. 2002a; Kaptein et al. 2006), illness perceptions, coping and quality of life of spouses (Helder et al. 2002b) and the role of patient and partner's illness perceptions in quality of life (Kaptein et al. 2007).

The SRM provides a framework to facilitate understanding of human behaviour in response to illness or stressful experiences. A major focus of the research by Levanthal and colleagues was to produce educational interventions that enhanced people's understanding and behaviour with regard to health practices. Levanthal et al.

(1984) hoped that the SRM would facilitate development of programmes for promoting and maintaining changes in health practices. They were particularly interested in the application of the model to long term conditions such as hypertension and diabetes. The model has since been used to explore illness perceptions in various conditions including and not limited to Huntington's disease, beliefs about Cancer (Llewellyn et al. 2007), Chronic Fatigue syndrome (Moss-Morris et al. 1996), Multiple Sclerosis (MS), (Vaughan et al. 2003), Rheumatoid Arthritis (RA) (Hill et al. 2007).

The model is depicted diagrammatically as a feedback 'loop' (see figure 5). Illness representations form a vital component of the model and form the main focus of the research using the SRM in HD. Illness representations are formed from different experiences, beliefs and social and cultural context. Put simply, self-regulation is a processing system where individual's move through stages of information from when a health threat is received, representations of the threat are formed, acted upon (problem solving, goal planning, action plan), evaluated and then perceptions are integrated into memory. Illness representations (cognitive representations of the illness threat) guide coping responses and actions, which are then appraised. The appraisal is then fed back and integrated into memory to further inform the representation and the individual may subsequently modify their behaviour or action based on the perceived success of the behaviour. Appraisal may also lead to confirmation of the use of certain strategies or actions and consequently possible reappraisal of their illness representations and so the cycle continues. The model is recursive and so this is an ongoing cycle where representations alter or stay the same through subsequent 'episodes'.

Figure 5: The Self-regulation Model (adapted from Levanthal et al. 1984)



Levanthal et al. (1984) describe four basic assumptions specific to the SRM. The first is that of 'active processing'. The SRM assumes that an underlying information processing system integrates current information received ('stimulus information') with innate and acquired memories, which influence the construction of behaviour and experience in an episodic fashion. i.e. on a moment by moment basis, the processing system uses the information to create the emotional and coping reaction to the world around us. The second assumption relates to 'parallel processing', i.e. the processing system is divided into parallel pathways. As an individual adapts to a specific situation, the pathways interact, with one developing an objective view of the illness threat/representation and the other, an emotional response to the situation. Following creation of an objective view, a coping plan for managing the threat occurs, whilst a plan for the management of emotion is also created in reaction to the emotional response. In addition, objective information about attributes of the representation may affect emotional reactions which can intensify or diminish symptomatology affecting the representation and alter the coping response.

The third assumption of the model is that of 'stages in processing'. The SRM suggests that the processing system operates in stages and that they are recursive. The first stage is described as creating the representation of the problem and the accompanying emotion. In the second stage response plans for coping with the problem and the emotion are developed and carried out. The representation guides the coping stage; the representation or 'problem' as it could be described, drives the goals for coping. During the third stage, appraisal of the coping response is carried out to determine whether the individual has gotten closer to the coping goals set in response to the representation or not. The information obtained from the appraisal stage feeds back into the prior stages. The coping strategies employed and or perceptions of the representation may then be altered. The underlying memory structures are altered with each cycle of the process or 'episode' which thereby influences and changes subsequent cycles of the process or 'adaptive episodes'. This means that information is processed iteratively and repeatedly cycled through the stages described, (representation to coping to appraisal).

The fourth assumption is defined as 'hierarchical processing'; the processing system is hierarchically organised, operating at both concrete and abstract levels. Appraisal at one level (e.g. appraisal of a coping response guided by an abstract concept) may feed in new information to the processing system and alter the information processed at the concrete level. Processing may begin at either level. For example, abstract information such as somebody being told they have a condition may lead to the individual checking

this hypothesis 'I have a condition' against the concrete experience; directing attention to physical sensations or symptoms. Once the individual confirms these for themselves, this information is integrated into the system and contributes to the formation of the illness representation. If they have previously experienced the condition, memories from previous episodes also contribute to the construct of the representation. Indeed, prior experience of a type of concrete information may lead to a quick labelling of the event/condition/symptoms and taking action in terms of self-treatment. An example is of indigestion, something which recurs and once the symptoms are recognised, the symptoms are labelled as relating to indigestion and non-prescription medication is taken as the action plan/coping response. An example given by Levanthal et al. (1984) of the two different levels working is that of coping with a headache. They suggest that in the behavioural episodes of coping with a headache, the concrete features are the pains associated with a headache and the abstract feature is maybe the idea that one has had a stroke. This suggests the possibility for there to be discrepancy between the concrete and abstract levels created by the hierarchical component of the processing system. Another example of inconsistency between the two levels given by Levanthal et al. (1984) is of a patient agreeing to and going through a medical treatment to reduce the size of a tumour. The consultant may give them positive results that the treatment worked but they feel worse during and after treatment than before commencing the treatment. Levanthal et al. (1984) explain that whereas emotional responses are more likely to be dependent on concrete processing and perceptual memories, abstract information is more influential on the creation of problem based representations.

From their studies of cancer treatment (Nerenz 1979), Levanthal et al. (1984) saw how inconsistencies in the regulatory system can cause distress. Inconsistencies may occur regarding a change in the disease meaning that the individual's coping plan is no longer appropriate, or bodily sensations experienced that are not attributable to the current treatment somebody may be receiving. One can consider this in the context of HD and PA, where health care professionals may encourage activity as part of self-management from an early stage prior to manifestation of symptoms. Although the healthcare professional knows that this is part of the ongoing management to maintain abilities and perhaps delay onset of symptoms, if the individual with HD does not understand, they may become distressed because they do not understand the reason for the treatment when they have no symptoms.

The model describes the attributes that construct illness representations as five different domains: 1) Identity; symptoms or names given to the disease; 2) consequences; symptoms and impact on life functions; 3) cause; whether the cause is

infection or genes, internal or external; 4) control; disease perceived as preventable, curable or controllable; 5) timeline; expected duration or age of onset.

These representations created (as described earlier) can be abstract or concrete, but integration of the two levels of information into a consistent whole is an important step in constructing illness representations. Illness representations are developed from different sources of information including from an individual's personal experience of the illness, illness-related beliefs, and the social and cultural context. In the wider cultural context this may be for example, information about illness from the medical care received, in the media. In the more personal context this is information through direct social communication with others, which may be family, friends and health care professionals for example. To illustrate this, Levanthal et al. (1984) suggest that symptom appraisal involves the sharing of information with those in their social network; friends, colleagues, family. Information sharing can lead to formulation and evaluation of hypotheses related to components that form the illness representation; cause, consequence, timeline (duration of illness), treatment options or coping plans and likely outcomes of those. From these interactions a construct of the problem is created leading to the plan of action (self-medicate / adopt a health behaviour / seek medical advice). If the plan includes to seek medical advice, the individual is then exposed to the knowledge, beliefs and expectations of the healthcare professional which will also impact on the individual's illness representations.

Levanthal et al. (1984) discuss the interaction of objective representation and emotion. Emotions can alter response to illness in a number of ways through influencing the representation of the illness. Emotion can affect the amount of energy available to take action in response to illness, and the perceived intensity of the illness that can be mistakenly attributed to the illness. This is argued in that illnesses and emotions can both affect bodily sensations such as stomach activity, irritability and heartbeat (Levanthal et al. 1984). Therefore, emotion can influence illness representations and feedback to either reinforce or challenge the value of coping responses. From another perspective, illness can have complex effects on emotion. Components of the individual's representation of the illness (identity, timeline/perceived duration, perceived consequences) can produce affective reactions generated by either the abstract (e.g. cancer, stroke) or concrete labels (lump, impaired function) of the representation. Combinations of these, such as perceived extended timeline of severe consequences, may have potent effects in provoking emotions. For example, chronic conditions generate emotions of despair and depression (which may be applicable to HD), as the

duration of the illness duration is essentially for their lifetime which may be shortened, and the consequences severe.

Understanding how people perceive and manage HD could help gain better understanding of how the nuances of HD influence PA experiences and engagement. In terms of the relevance of the SRM for HD, it appears that the SRM could be a useful model to help further explore the impact of HD on PA experiences. The illness representations of timeline and identity seem particularly pertinent considering the issues surrounding identity in HD and changes with progression of HD elicited in the earlier sections. The concept of coping has been elicited as a major part of life with HD (Helder et al. 2002; Kaptein et al. 2006; Aubeeluck et al. 2012) (see appendices 2 and 4). Exploring how PA might fit into this aspect of HD could help understanding of experiences and perceptions of PA in HD. It seems that a non-linear model to support exploration of PA experiences would be appropriate as people with HD are always adapting in response to HD progression. People potentially go through ongoing reappraisal of how they cope with certain situations or have to change behaviours to accommodate their changing abilities. This could be relevant for PA as the progressively worsening symptoms have implications for what people are able to do and hence the representations linked to the somatic experiences of the body guide and influence PA.

Qualitative research has demonstrated the relevance of SRM for helping to understand experiences of other neurodegenerative conditions. In addition to the studies in HD, the SRM has been used in qualitative research to assist in exploring the experience of developing dementia (Clare & Harman 2006). Semi-structured interviews with people with dementia were analysed in two stages. Open coding was firstly used to identify themes followed by a theory driven content analysis where they coded elements of participants' illness representations under the categories of identity, timeline, causes, consequences, and control (Clare and Harman 2006). Using the illness representations as a framework for analysis as well as open coding of the data allowed the authors to capture insight into experiences of dementia and how people negotiate day to day life with early stage dementia. The SRM contributed to furthering understanding of the personal dilemmas that people face with progressive decline and therefore provide insight into how individuals might be supported to deal with them. The SRM is therefore appropriate to use in the aim of understanding how people manage onset of dementia and the ongoing experience of it.

Considering that progressive dementia is part of the pathology of HD (Peavy et al. 2010), application of the SRM is appropriate in the context of HD to gain insight into the

impact of it on PA engagement. Matchwick et al. (2014) used the SRM as a theoretical underpinning to explore the illness representations of cause and control of individuals with Alzheimer's disease (AD) through conducting semi-structured interviews.

Matchwick et al. (2014) found that participants made sense of and developed an understanding of AD through using prior experience of physical health illnesses. Again as AD has parallels with HD, it could be interesting to explore how past experiences of PA before onset of HD inform how they make sense of engaging in PA with HD.

Following evaluation of the theoretical models used by the studies identified for the literature review, the SRM is proposed to be an appropriate model with which to further understanding of experiences of HD and how the nuances of HD impacts on PA engagement and experiences. In summary, the SRM is favoured over the other models reviewed (WHO-ICF, SCT and TMSC) because of the inherent linear nature (SCT and TMSC) and lack of utility to account for change over time (WHO-ICF) which may make them less relevant for exploring experiences of PA in HD because of the complexities, and multidimensional and progressive nature of HD. Previous studies exploring experiences of PA in HD have lacked theoretical underpinning and so use of the SRM to underpin this research will contribute novel insight and a novel approach to exploring this area.

### **3.7. Chapter summary**

The literature review has guided the researcher's thinking about the methodology and research questions. A clear gap has been identified in the knowledge of personal experiences of PA in HD. There is a lack of stand-alone qualitative studies that seek to understand the impact of HD on PA participation or the experiences of people with HD in trying to engage in PA from their point of view. The changes that people experience over the life cycle of the disease could have implications for how PA is experienced and sustained have yet to be explored and the self-regulation model appears to be a relevant model that could facilitate this exploration. A study which explored the experiences of living with dementia used the 'illness representations' components of the SRM as a framework to analyse qualitative data. This provided a transparent way of gaining insight into the participant's experiences through the 'lens' or perspective of 'illness representations'. The studies that exist in relation to experiences of PA were all conducted as part of process evaluation of specific exercise or rehabilitation interventions, lack robust theoretical underpinning or are not clear about how theory facilitated understanding of experiences. In addition to this, the need to support people with HD to engage in PA was identified but a lack of supportive resources was

identified. It appears to be important to elicit perceptions of caregivers of people with HD because of the vital role that they play in the daily life of the person with HD. What is different about HD in comparison to PD for example is that onset can be much earlier in life. Spousal caregivers therefore may be working full time in addition to bringing up young children and caring for their spouse. Their time is precious, insight into the day to day living with HD may be helpful in improving understanding of how to support them to support the person with HD. With these considerations in mind the main research question, aims of the research and methodology were developed.

**Research Aim:**

Explore how living with HD impacts on the experience of physical activity participation.

**Research Questions:**

- 1) What are the experiences of physical activity participation across the stages of HD from prodromal to late stage?
- 2) How do the nuances of living with a neurodegenerative disease such as HD affect engagement in physical activity?

This research aimed to address the gap in knowledge by exploring perspectives and experiences of physical activity in HD and contribute to the limited body of evidence regarding physical activity experiences of people with HD. Further, the findings could be informative to the development of complex interventions of exercise in HD, clinically and in research studies, in considering the contextual factors and influences (Craig et al. 2008; Moore et al. 2015). A qualitative methodology was developed that utilised the self-regulation model to facilitate exploration of how living with HD impacts on the experiences of PA participation, to answer the research questions.

## **4. Methodology**

The following chapter describes research design development regarding sample, data collection, data analysis and ethical considerations. To address the aims, the target sample was people who had an interest in discussing physical activity (PA) and were either gene positive or had Huntington's disease (HD), and/ or were caregivers and/or family members. The sample and research method rationale are discussed in the following sections. To give a brief overview, the focus group (FG) method was used to explore experiences and perceptions about PA of people with HD and their caregivers and/or family members. FGs were conducted as part of the ENGAGE-HD trial intervention development workshops (see appendix 7). FG participants were people with HD across the stages of the disease and their caregivers and family members across the UK recruited through the Huntington's Disease Association (HDA).

### **4.1 The researcher's perspective; 'my perspective'**

The experiences gained from delivering the exercise intervention for COMMET-HD, and the findings of the literature review highlight the physical, bodily experience of HD and the importance of their social environment, mainly in terms of their familial caregivers and healthcare. Social and environmental interactions form a key part of the illness experience and the perceptions of the illness in HD (Downing et al. 2010; Skirton et al. 2010; Kaptein et al. 2007; Williams et al. 2007). My personal perspective aligns most closely with the post-modern social constructivist perspective which allows the researcher to focus on the worlds of the participants. It considers how participants construct their realities and the social, environmental and cultural influences on how they construct their life worlds and experience HD in addition to the bodily experiences of HD (Yilmaz 2007). My belief is that knowledge and experiences are not constructed in a vacuum and indeed individual existence is embedded within a complex set of social structures. The social constructivist perspective influenced the development of the research design and the decision to use FGs as this would allow me to explore the discussions and experiences shared and constructed by participants. The FG setting would allow participants to share their experiences and possibly gain a better understanding and explanation of their experiences through discussing them with other participants. i.e. leading to new constructed knowledge through the social participation with other FG members. This in turn could facilitate obtaining richer data and understanding as participants seek more understanding from each other and explore their experiences more in depth.

The past experiences I have had and complexities of social structures within my own world will have shaped my thought processes and understanding in all aspects of my life. In the context of this research it was my experiences as a physiotherapist and somebody who has worked closely with people with HD specifically in an exercise context that influenced the creation and development of this project. This is important to acknowledge, since my experiences may have shaped specific opinions or already formed ideas about how HD impacts on PA. Husserlian phenomenology discusses the ‘bracketing’ of thoughts or preconceptions to prevent their influence on the research (Hamill 2010). Although it is impossible to completely ignore those experiences, in acknowledging and recognising their influence at the outset of this research, I hoped to portray transparency of the research. I set about achieving this by defining a transparent method and describing the process of analysis followed where the focus is on describing and staying close to the data itself and not my own abstract thoughts.

From the outset of this research I personally acknowledged that the researcher needs to consider the potential impact of their own personal perspective, experiences and values on the research they carry out. Indeed, demonstration of this thought process is essential for qualitative researchers to portray rigour and trustworthiness of such work. The opposite to this, which is the consideration of the research impacting on the researcher is also something that I acknowledged and recognised as a possibility at the beginning of the process. Yet it was only through my experiences of data collection and analysis that I began to fully appreciate the meaning of this to me. During data collection and analysis, the participant responses and findings caused me to return to reflect upon my own experiences of delivering the exercise programmes in the HD exercise studies using reflective diary notes. This gave me a greater appreciation of the potential issues that participants had faced without me even being aware. Although it is important to acknowledge that only the people living with HD can ever truly experience how HD affects PA participation, sharing their insight can help others to understand. The opinions and experiences of participants certainly helped my personal understanding of HD and in the process reshaped some of my thoughts about PA in the context of a neurodegenerative disease.

## **4.2 Research design and rationale**

An approach that enables interactions to facilitate exploration of individuals' constructed perceptions and experiences about PA in the context of HD was required to enable exploration of how the impact of living with HD impacts on PA experiences across the stages. The wider culture of participants' societies brings with it its own

values and impact on how they make sense of and attach meaning to experiences in their social world. However, in a sense the researcher was accessing a smaller culture within the wider culture where HD is the main factor which impacts on the participants' lives (particularly PA for this research) and also affects construction of their own reality and perceptions. The FG method is conducive to facilitating exploration of such constructs of perceptions and experiences. In light of limitations discussed in the introduction and literature review regarding the individual interviews of the COMMET-HD RCT that the researcher analysed (Busse et al. 2013), the FG method would allow for the wider perspective of the family members and or caregivers. HD is a family condition and family members and caregivers may bring a slightly different insight that would be valuable to capture. Discussed in more detail in section 4.3.2. ('participants and recruitment') caregivers may help the person with HD to remember or identify issues that they may not elucidate (Carlozzi and Tulsky 2013). In addition, they may be able to explain how the person with HD has reacted in different situations and provide details, which the person with HD may have forgotten (Clare and Harman (2006).

### **4.3 Focus groups**

Focus groups (FG) are a useful way to uncover factors that influence behaviours, motivations and opinions (Casey MA 2009). Interactions also highlight participants' views of the world and enable them to ask questions of each other and potentially re-evaluate and re-consider their own understandings of their experiences, (Kitzinger 1994; Kitzinger 1995). FGs were chosen as the data collection method in consideration of the research aims and the researcher's previous experiences of individual interviews with people with HD. In addition, the FG method has previously been useful in exploring experiences of HD; FGs with family members and caregivers, FGs which grouped together people with HD at similar stage, and FGs which grouped people together across the stages of HD (Williams et al. 2007; Hartelius et al. 2010; Carlozzi & Tulsky 2013). FGs allow the researcher to observe and gather rich data pertaining to the understanding, sharing and sense making of experiences when participants of the groups come together.

Within FGs there are opportunities for participants with common issues to share insights during a focused discussion whilst also providing diversity of perceptions and experiences (Krueger & Casey 2009). Being able to obtain diversity within the FGs is important because of the research aim of developing understanding of PA experiences across the stages of HD. As FGs allow for inclusion of a larger number of participants, this data collection method facilitated meeting the aim of obtaining experiences across the stages of HD and allowed for inclusion of caregivers to explore their perspectives

and experiences too, in perhaps a less formal situation than if it was just the caregiver and person with HD being interviewed by the researcher. Being able to collect large volumes of data over a relatively shorter time period compared to interviews is an additional advantage of FGs. Multiple voices can be heard at one sitting of a FG giving rise to a larger sample from a smaller number of data collection sessions (Kitzinger 1995).

A limitation of the individual interviews of the RCT (Busse et al. 2013) (discussed in chapter 3; literature review) was that often only one word answers were given or else very short answers to the questions even with prompting. Observation of this also informed the decision to use FGs. The dynamics within a group of people facilitate sharing of ideas and experiences that possibly may not be brought up in an individual interview. In comparison to individual interviews, FGs have a different dynamic where the discussions within a group may lead to exploring unanticipated avenues of thought (Krueger & Casey 2009). Considering the potential challenges of conducting FGs is important, especially in this context where there may be participants whose ability to communicate as well as others may be compromised due to HD symptoms. Hartelius et al. (2010) decided to conduct FGs that separated people with HD from the caregivers and family members, their rationale being that mixed groups may make the participants feel less free and inclines to speak about personal thoughts and feelings. Potential challenges and how these were considered and addressed are discussed in more detail in the section further on titled ‘Conduct of the focus groups: specific considerations related to participants with HD’.

#### **4.3.1. Development of the focus group schedule**

In developing a schedule of questions for FGs (appendix 12), Krueger and Casey (2009) suggest that questions that are easy to say and understand, are open ended, clear, short and conversational are the best to facilitate conversation. Questions developed with these key aspects in mind can help create a more informal sociable environment to help participants feel more comfortable. The questions in the schedule were developed to address the research questions. The schedule structure was developed following Krueger and Casey’s suggestion for a ‘questioning route’ (Krueger & Casey 2009). The questions themselves were developed using the findings of the reviewed literature that identified key features of the experiences of living with HD and PA experiences of HD and neurodegenerative conditions. These key features included aspects of progression of HD, the salient concept of motivation, strategies for engagement and barriers and facilitators to PA and illness identity (Helder et al. 2002; Kaptein et al. 2006; Zinzi et al. 2009; Elsworth et al. 2009; Ravenek & Schneider 2009;

Quinn et al. 2010; Debono et al. 2012; Ellis et al. 2011; Eriksson et al. 2013; Frich et al. 2014).

The schedule followed a questioning route (Krueger & Casey 2009) that started with an opening question to put people at ease to speak. This was followed by introductory questions that set the scene, the transition question that moved the conversation to the key questions, then the key questions which drive the study and finally the ending question to bring the discussion to a close. Detailed development of the schedule can be seen in appendix 12. The final schedule can be seen in appendix 13.

#### **4.3.2. Participants and recruitment**

##### ***4.3.2.1. Sample***

The impact of HD is experienced by both the people with HD and those close to them; family members (who may be caregivers), and formal caregivers have much insight from their work (Maxted et al. 2014). Given this, it was important to the researcher that caregivers and family members as well as people with HD were included in the FGs. Although HD has been portrayed as a socially isolating disease outside of the family unit, the increasingly integral role that caregivers play in the lives of people with HD places them well to understand the impact of it on PA (Williams et al. 2007; Aubeeluck et al. 2012; Maxted et al. 2014; Frich et al. 2014). Observing the dynamics between caregivers and people with HD may paint a more accurate picture of the real-life at home situation rather than if only people with HD were included (Maxted et al. 2014). Other than the people with HD, these are people who can have a role in supporting and encouraging PA participation as they have an integral role in the daily routine of the person with HD (Khalil et al. 2012). Therefore, caregiver's own beliefs and attitudes towards exercise can impact on PA participation of the person they care for.

It was also appropriate for caregivers to be present for those with HD at later stages who may need prompting or help in getting their point across to the group. Indeed, Carlozzi and Tulsky (2013) have previously conducted FGs with people across the disease spectrum and their caregivers. They cite involving caregivers because of the potential cognitive impairments that people may have had and would be recognised by caregivers who might help them to remember or identify issues that the patients themselves may not elucidate (Carlozzi and Tulsky 2013). Clare and Harman (2006) also suggest inclusion of the caregivers or family members may be of value since they might be able to explain how the person with HD reacts in different situations and provide details, which the person with HD may have forgotten.

#### *4.3.2.2. Recruitment Process*

The Huntington's disease Association (HDA) is a charity that supports families affected by HD. The HDA provides information and advice to professionals whose task it is to support HD families. The HDA employ Specialist HD Advisors (SHDAs) to provide a wide range of services including providing information and advice to families, answering crises calls, liaising with other professional service providers, liaising with local branches and self-help groups, giving talks and organising seminars and training days.

The SHDAs are based in different areas of the UK and have contact lists of HD families. The HDA maintain a confidential mailing list for members of the HDA who have agreed to be used for postal contact allowing them to approach people about taking part in research studies. The head office of the HDA was contacted to request information to be sent to SHDAs to see who would be interested in helping to run workshops where people with HD and their caregivers were invited to attend and discuss PA and exercise experiences. SHDAs responded from numerous areas around the country offering to help run the workshops. The workshops were conducted on the basis of positive responses from the SHDAs and geographical spread (Patton 1990). Common patterns or themes that emerged from variation were of particular interest and value in capturing the core experiences and central, shared experiences, in this case, of PA.

Information about the workshops was circulated to everybody on the SHDAs' databases in each region by post. All correspondence was initiated by the HDA and no personally identifiable details were provided to the researcher without consent of the involved individuals. The information specified that up to 15 people were invited who would be able to attend a group in their local area. It specified people who were interested in and able to participate in a group discussion about their experiences and views of exercise. Due to the difficulties often experienced in reaching the HD population to take part in research each contact on the HDA care advisors' lists was invited with the expectation of a low response rate. The groups were also advertised as being on a first come first serve basis in case of a large response. Where more than seven people attended, the group was split into two unless in exceptional circumstances where this was not possible (as was the case with the first FG). Challenges of conducting research in HD meant there was possibility that people registered and did not turn up on the day. Behaviour can be unpredictable in HD and also symptoms can vary meaning some might not have felt able to attend due to their symptoms on the day.

Research participants were therefore people gene positive for HD or had symptoms of HD and/ or family members and or caregivers of people with HD that received information about the study and were interested in discussing exercise and exercise experiences in a group format. They also had to be available at the date and time of the FG session in their area.

People at end-stage HD were not included in the research sample. As discussed in the literature review, people at this stage are bed-bound and severely impaired in terms of function, cognition and communication. Therefore, it was not appropriate to expect to recruit participants to FGs at end-stage HD.

#### *4.3.2.3. Conduct of the focus groups: specific considerations related to participants with HD*

Due to the nature of HD and difficulties some people may have with their speech or initiating conversation (Saldert et al. 2010) there was potential for the issue that not all voices would be heard equally. This was acknowledged but would not be known until the FGs took place. For some people with speech difficulties their caregivers asked for permission from the person with HD to speak on their behalf and in some cases clarify what the person was trying to say. The caregiver presence, particularly for those with late stage HD could help ameliorate the possibility of memory and communication deficits affecting the patients' recall and expression of experiences (Walker 2007). This approach has previously been used with stroke patients who may have had similar issues (Lilley et al. 2003). In order to include people with late-stage HD this was something that had to be accepted in the case of some participants. However, everything the caregivers said was checked with the person with HD for a nod or another indication such as hand gesture that there was agreement with what the caregiver had said (Lloyd et al. 2006). One reason for using the FG method rather than individual interviews was that people with HD may struggle to initiate answers to questions due to the cognitive impairments. The researcher considered that a FG format and dynamics within a group setting meant that anybody with HD who may struggle with such issues would not necessarily have to initiate discussion. Once discussion had begun it would be easier to maintain a flow as people bounce ideas off each other generating more discussion (Krueger & Casey 2009).

FGs have previously been described as 'empowering' participants in terms of talking with other people with similar experiences (Kitzinger 1995). In a disease with no cure it was possible that participating may have empowered participants to feel that they were contributing something to developing understanding and research in this area.

Alternatively, by inviting people across the spectrum of HD to the FGs there was a potential issue of participants being upset by seeing others at later stages of the disease. The researcher made it explicit to attendees that they were free to leave at any point without giving a reason and that their local SHDA was present if they wished to seek advice. The local SHDA attended each FG. Having been the ones to make direct contact with the participants about the FGs, the SHDAs had previously been in contact with participants and had known most of them over a period of time. The SHDAs participated in the FGs but were also present in case of any issues or if participants needed advice. So that the SHDAs did not lead any participants in their answers (as some were familiar with the participants) or take over the discussion, the researcher clearly explained the aims of the research and ground rules including that everybody should have equal opportunity to speak for themselves and everybody's opinion or experience is equally important (described in the 'subsequent data collection' section). All participants were made aware that the SHDA was there to support them. The organisation of the FG sessions was such that participants had time for refreshments and informal conversations before the sit down discussions began. The context of HD provided the common ground for those who attended and before the FGs proper had started, time for refreshments and conversation facilitated building of rapport between participants.

## 4.4 Data collection

According to Casey and Krueger (2009) three to four FGs should be conducted after which it should be decided whether saturation has been reached. A pilot FG session was conducted to trial the method and the schedule. To ensure inclusion of all stages of HD seven further FGs were conducted in different areas of the UK. In organising the FGs disability access was ensured at all locations.

### 4.4.1. Focus Group 1: Pilot Group

Due to circumstances outside of the researcher's control there was only one room available rather than two (as had been originally organised). Considering people had used their spare time to attend, the researcher felt that it would not be ethical to send some people away to achieve a smaller group. Alternatively, conducting 2 FGs one after another was not feasible in terms of time. With 15 people in attendance and no option to split the group it was challenging for a number of reasons. Some break out discussions between different participants took place simultaneously and capturing everything that was said was difficult when transcribing. Also having such a large group, it was possible that some would have felt less comfortable speaking in front of the others, especially if they had speech impairment or lacked confidence. Another

limitation could have been that due to the large group, it was possible that not everybody had the chance to say everything that they wanted to. Some of the participants knew each other, which, it seemed to help relax the atmosphere but others may have felt more self-conscious because of this. Nevertheless, because it was not possible to split the group there was more than one facilitator. This meant that if more than one person was speaking it was easier to manage the group and was possible to capture interesting perspectives and discussions that took place. In terms of moving forward to the next FGs, this emphasised the importance of an appropriate environment in which to conduct the FGs.

#### *4.4.2. Subsequent Data Collection*

As FGs are conducted, it may be found that some questions are inappropriate in the sense that there is no response or participants do not understand. Although a pilot group was conducted to test the questions, questions were sometimes slightly adapted for the understanding of participants (Krueger & Casey 2009). At the beginning of each FG, the participants were welcomed, given an overview of the topic for discussion and the researcher's role as facilitator. It was explained to the group that participation was voluntary, and participants were able to withdraw at any time without giving any reason, and it would not affect their medical care or legal rights in any way (Morgan 1996). The researcher advised participants that conversations would be digitally tape recorded but in all transcripts participants would have a pseudonym so would not be identifiable. The participants were then given a chance to ask questions about the study before reading and signing the consent form (appendix 10). Participants were informed that the FG would take approximately one hour.

At this point the researcher's role as the facilitator was explained and the ground rules were introduced. These were: only one person speaks at a time; confidentiality is assured "what is shared in the room stays in the room"; it is important to hear everyone's ideas and opinions, everybody has the right to be heard equally and we may sometimes ask you to expand on something you have said; there are no right or wrong answers to questions – just ideas, experiences and opinions, which are all valuable and it is important for us to hear all sides of an issue – both the positive and the negative (Krueger & Casey 2009). Notes were recorded by an assistant moderator who dealt with any unexpected interruptions, assisted with environmental conditions and logistics and checked the tape recorder. During the FGs it was not uncommon for participants to encourage others who were trying to share their experiences but had speech difficulty or were quieter than others. This kind of facilitation may have been less intimidating than if the researcher facilitator was to do more of the probing as an

outsider i.e. somebody who has not lived with HD. As well as sharing their experiences, participants bounced ideas off each other, asking for each other's opinions which facilitated more in depth discussion around issues.

Once the discussion for question four had reached saturation, a brief summary of the discussion was given and the group was asked whether it was an adequate summary in their view. This gave participants an opportunity to reaffirm anything poignant to the group, reflect on previous comments and also clarify their position if they realised that they had made conflicting comments during the discussions. This final question also acted as an insurance for the researcher, to ensure critical aspects were not overlooked (Krueger & Casey 2009).

The researcher's awareness of the effects they may have had on the FG dynamics allowed them to ensure that they did not respond or react in ways that showed bias during the interactions of the FGs that may have influenced people's discussion and imparting of their perceptions and experiences. For every FG, at least one other physiotherapist experienced in working with people with HD also attended. This was because until the sessions took place it was not known exactly how many people would turn up to take part, given the unpredictability of behaviour due to symptoms such as apathy, or cognitive impairment that affects memory (Marder et al. 2000; Naarding et al. 2009; Reedeker et al. 2011; Thompson et al. 2012). If a FG had to be split to form two FGs because of large numbers, there was therefore another physiotherapist experienced in working with people with HD available to facilitate another other group at the same time. It is possible that the other facilitators had different ideas of interesting avenues to further explore and ask participants to expand upon. The researcher attempted to limit this by discussing their research aim explicitly with the other physiotherapist and the research questions they were hoping to answer for the study. Demographic data was collected via a short questionnaire to be able to describe each group and the people who took part in terms of their current PA, and level of assistance required, stage of disease (appendices 12, 13).

Each FG was grouped by convenience on the day as it could not be guaranteed how many participants were going to turn up given that people with HD can be prone to having unpredictable 'good' or 'bad' days. Where larger numbers attended the data collection sessions, people naturally drifted into groups and separated into people at different stages during the refreshment sessions before the formal session. The researcher was aware this may happen due to the progressive nature of the disease. In addition, people with MS and PD preferred being with those at a similar stage in

support and exercise groups (Ravenek & Schneider 2009; Elsworth et al. 2009; Learmonth et al. 2013). The groups that had naturally formed were left as they were. The natural forming of the groups suggested that the participants were comfortable with each other and had begun to develop interactions which may have been more supportive of flowing discussion during the FG session. FGs were conducted over a few months to allow time between them for reflection of each group and whether the schedule needed adaptation.

## 4.5. Data analysis

### 4.5.1. Overview

An adapted version of the framework method of analysis (FA) developed by Ritchie and Spencer (1994) was used for analysis of the FG data. FA provides transparency, facilitating a clearly defined process that can be replicated. The method enabled the researcher to systematically develop key themes which describe and map the data. The five stages of FA are: familiarisation with the data, developing and refining an analytical framework, indexing the data using the analytical framework, charting the data into matrices, and mapping and describing (discussed in the following sections of this chapter).

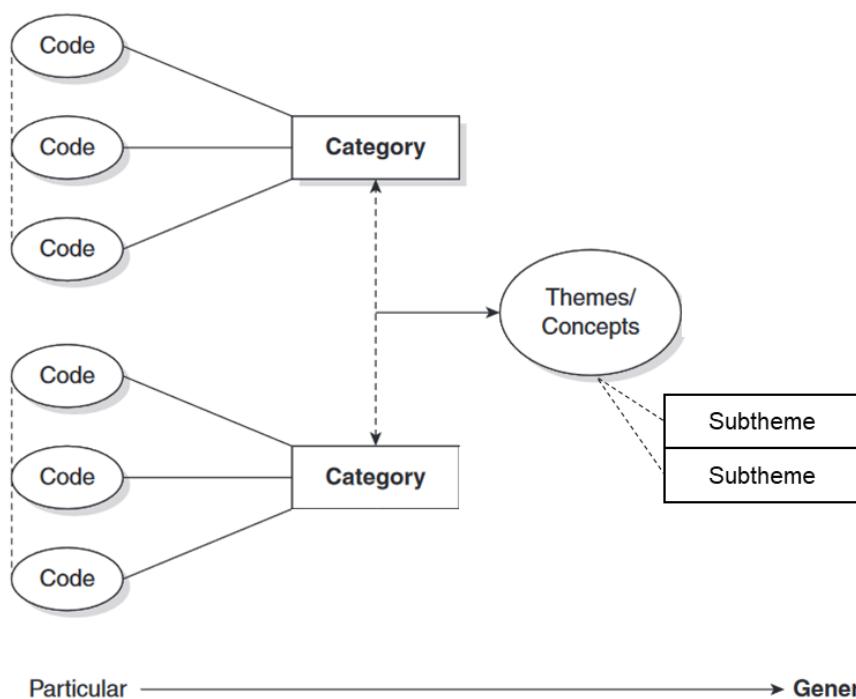
As each of the stages is well defined in FA, this method also allowed the researcher to revisit earlier analytic stages. For example, after indexing and charting the data using the analytical framework, the researcher periodically went back to review the memos made while indexing to help consolidate their thoughts. Memos were used throughout the analysis process to help the researcher capture thoughts about the data when coding and to keep track of decision making about development of key themes (Birks et al. 2008). Memos were usually hand written notes that were kept in a ‘reflective notebook’ but NVivo also allowed for notes to be made when coding the data, called ‘annotations’ so some notes were also entered into NVivo (example in appendix). An example of when the researcher used a memo to consolidate thoughts was when the researcher returned to a memo they had written about ‘identity’ when coding the data initially. Reading the memo (appendix 21), led the researcher to reread the transcripts to confirm their thoughts about the context in which identity was identified as a salient issue. Doing this helped to consolidate the researcher’s thoughts about how and why ‘identity’ was coming across as a strong thread and with the decision to make it a sub theme of key theme 1 (“the evolving representations of Huntington’s disease and their impact on physical activity”). As demonstrated by this example, following the thought patterns of the researcher, analysis was not always a linear process. Although the transcripts were mostly read during the ‘familiarisation’ stage, the researcher also

returned to transcripts much later in the analysis process too. This was to confirm or negate thoughts and check that the developing themes and subthemes truly represented what the participants had shared.

Although other research using FA has analysed the first transcript and used the themes derived from that to drive the line of inquiry for the next interview schedule, (Swallow et al. 2015), due to logistics of when the FGs could be conducted, the researcher did not use this approach. However, following each FG, the researcher reflected on the appropriateness of the schedule and discussions that had occurred during each FG. Following the pilot FG, it was found necessary to adjust the introductory question (as described in section 4.3.1.).

A key part of FA is development and refinement of the analytical framework used to index the data, which is discussed in the following sections. The initial framework was tested by the researcher and two other individuals (described in a later section) by applying it to the data. Following this activity and discussion about the framework it was refined to form the final analytical framework to use in the subsequent steps of data analysis, which led to development of key themes described in sections 4.5.2.6. Essentially data analysis led the researcher to move from codes to categories to themes as depicted in figure 6.

Figure 6: Example of developing themes adapted from Saldaña (2009)



#### *4.5.2. The analytical framework*

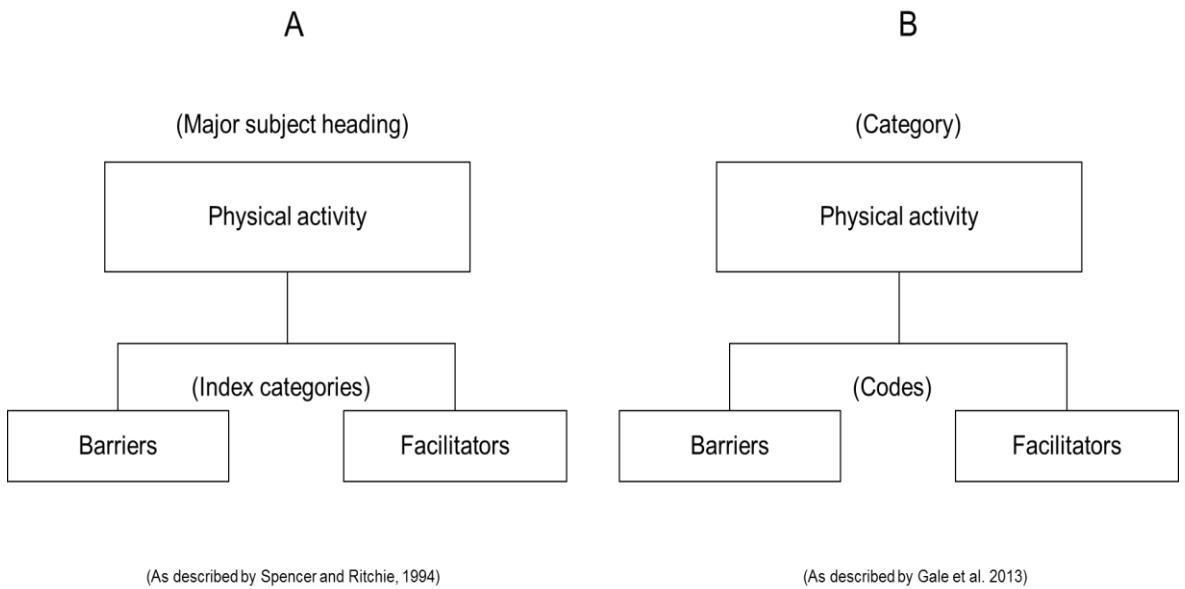
The analytical framework described by Spencer and Richie (1994) is composed of major subject headings, which are made up of index categories. For example, a major subject heading may be ‘physical activity’ with index categories of ‘facilitators’ and ‘barriers’ (figure 7) Gale et al. (2013) name these differently, describing them as ‘codes’ and ‘categories’. Codes can be grouped together to form categories that are clearly defined, in the same way that index categories described by Richie and Spencer (1994) can be grouped to form major subject headings.

In qualitative data analysis, codes can be grouped together to form categories, which are compared and consolidated to progress towards and form themes that transcend the specific data itself represent the data overall (Saldaña 2009). Considering this process of analysis, of moving from codes, to categories, to themes, to the researcher’s thinking, logical terminology would be to use the words codes and categories to describe the framework, rather than index categories and major subject headings. In other words, codes and categories form the analytical framework. Consolidation and comparison of those categories (through indexing the data using the analytical framework, followed by charting and mapping of the data) would then form the descriptive key themes of the findings.

Using the terms ‘codes’ and ‘categories’ also seemed logical because the analytical framework is developed partly from codes identified through open coding of the data. Following open coding of the transcripts (described in more detail later in this chapter), these codes are reviewed to decide which ones form the codes of the analytical framework. There is a step where the researcher decides which of those codes will be included in the analytical framework (between identifying codes through open coding and putting them into the framework). From the researcher’s perspective, these initial codes do not yet form ‘categories’ as they are essentially the phrases or words that capture the first impressions of the data, which are organised and grouped into categories in a subsequent step of the analysis. Therefore, the researcher would have found it difficult to justify why at this point, they were being called ‘index categories’. The label of ‘codes’ seemed to fit better and the researcher wanted to use the most transparent terminology to be able to clearly describe and justify the process of analysis followed.

Figure 7 depicts the different terminologies used by Spencer and Ritchie (1994) and Gale et al. (2013) to describe the components of an analytical framework. As discussed, the researcher used the terminology used by Gale et al. 2013 (diagram B).

Figure 7: Comparison of terminology for analytical framework components (adapted from Spencer and Ritchie, 1994; Gale et al. 2013).



The codes and categories that form the analytical framework can be developed either 1) through open coding of the data (inductively); 2) using existing theory, where the codes and categories of the analytical framework are predefined based on previous literature and or existing theories (deductively), or 3) using a combination of both. The researcher chose to use a combination of both, discussed below.

Previous research (discussed in the literature review chapter), aiming to understand the lived experience of dementia has used components of the SRM as a priori codes to conduct a theory driven analysis (Clare & Harman 2006). Theoretical underpinning in qualitative research gives transparency to the perspective from which the researcher is looking at the phenomena (Green 2014). The SRM was identified in the literature review as an appropriate model with which to underpin this research. In particular, the researcher was drawn to the use of a theoretical model to underpin data analysis. This was because of the novelty of such an approach in the context of HD and PA, where there is a lack of theoretically driven research that has aimed to explore general experiences of PA in the context of the complexities of HD. Illness representations in particular play a significant role in experiences of living with HD and perhaps would be relevant for PA in HD (Helder et al. 2002a; Helder et al. 2002b; Kaptein et al. 2007;

Kaptein et al. 2006; Arran et al. 2014). However, it was recognised that developing an analytical framework using only a priori codes created from the SRM may narrow and limit a full exploration of the data collected. In consideration of this, a combination of the two approaches was used (Gale et al. 2013) to incorporate a theoretically driven analysis using the Self-regulation model (SRM) but that was not restricted to only identifying data relevant to the constructs of the SRM. To achieve this, constructs of the SRM were used to develop a priori codes and categories of the analytical framework (Rashidian et al. 2008) and then open coding of all data was conducted to develop other codes and categories. Open coding all transcripts allowed the researcher to consider all of the data and make sure that none was overlooked (Ward et al. 2013). As this research was the first qualitative project to explore PA across the stages, it was important that all interesting aspects of the data in the context of HD and PA were explored.

#### *4.5.2.1. Development of codes and categories of the analytical framework: A priori codes and categories*

As mentioned in the literature review and previous section, the SRM was identified to use in driving part of the analysis. The SRM was therefore used to develop a priori codes and categories of the analytical framework. To ensure that the analytical framework captured the essence of the SRM, the components (described in the literature review) that contribute to the self-regulation process described by Levanthal et al. (1984) were added to the framework as the a priori categories (see table 2). These were namely, ‘identity’, ‘timeline’, ‘cause’, ‘control’, ‘consequences’, ‘coping’, ‘evaluation’ and ‘integration of perceptions with memory or experiences’.

Assigning constructs of the SRM as codes in the analytical framework as has been carried out previously in other research (Clare & Harman 2006). Although strictly speaking, ‘identity’, ‘timeline’, ‘cause’, ‘control’ and ‘consequences’ are sub components that form the overarching construct of ‘illness representations’, each of the illness representations are in themselves large topic areas that justify being included in the analytical framework as individual categories. Within each category, codes were added which were issues considered to be relevant to each category. These were identified by the researcher by reviewing the descriptions of the self-regulation model given by Levanthal et al. (1984), the Illness Perception Questionnaire (IPQ) which provides descriptions of the coping, control, consequences, identity and timeline (Moss-Morris et al. 2002) and also literature specific to HD that had explored and discussed illness representations (Helder et al. 2002; Kaptein et al. 2006; Arran et al. 2014). From these sources, descriptions for each code were developed to ensure that

at the indexing stage of analysis only relevant data was indexed to the analytical framework. However, to make the codes relevant to the topic of PA and less generic, the context of PA was added to the descriptions as a consideration. Table 2 shows the categories, codes and their descriptions. Once the initial a priori codes and categories were identified, the researcher moved onto developing the rest of the analytical framework using open coding. An important step before conducting open coding is familiarisation with the data.

Table 2: The a priori codes developed from the SRM

Category	Code	Code description
Coping	Strategies: physical activity	Specific strategies used to overcome limitations or barriers in relation to physical activity
	General strategies	General strategies of coping used
	Normality	What is normal to participants at different stages in terms of physical activity?
	Adapting	Adapting behaviour and or approach to physical activity because of the limitations caused by HD.
Evaluation	Reflection	Reflecting on specific experiences related to physical activity.
	Emotional experiences	Emotional responses to experiences – how do people react emotionally to certain experiences, what or how does this impact what they do in terms of PA?
	Objective response	How have people responded in a practical way to an experience they have reflected on – i.e. do they respond by troubleshooting, adapting what they do, how they do PA, ask or help or advice?
Cause	Lifestyle	Health behaviours - behaviours such as PA influencing health – what are people's perceptions of this, how does this influence their behaviour.
Consequences	Impact of symptoms	Impact of HD on physical, social, psychological function.
	Continuity	Perspective of how life will continue (also in terms of PA) in the context of the consequences of HD.
	Quality of life	Perspectives with regard to quality of life, how is it altered? Does PA mean anything in terms of improving or maintaining quality of life?
Control	Managing	Perceptions of treatment / management of HD.
	Healthcare	Contact with healthcare professionals.
	Health behaviour	Any perspectives of or beliefs in preventative health behaviours including PA.
	Control over PA	Meaning or perspectives associated with control when it comes to PA related behaviour.
Identity	Impressions of HD	How their impressions/ perceptions of the disease shape their attitudes/perceptions towards PA.
	Living with HD	How experiences of living with the disease shape their experiences and how they relate to PA for example – if they think it is not possible for them and it previously was- they lose that identity of someone who is physically active.
	Symptoms	How signs and symptoms of HD shape experiences of PA, meaning of the symptoms in the context of PA.
	Family	As a genetic disease – how family members deal with HD family history.
Timeline	Change over time	Anticipated change over time and progression of the disease.
Integration of perceptions with memory or experiences	Perceptions affected by experiences	Integration of perceptions with memory or experiences.

#### *4.5.2.2. Familiarisation*

The data was transcribed verbatim into Microsoft word. The researcher immersed themselves in the data by repeatedly listening to the audio recordings when transcribing. Pauses, emphasis of words, loud utterances, gestures and overlapping speech were indicated in the transcripts as suggested for FG transcription (Bloor et al. 2001). According to Bloor et al. (2001), a “thorough transcription of the tape recording of the FG is required for detailed and rigorous analysis to take place.” Every effort was made to transcribe all recorded speech, including when more than one person was speaking, unfinished and interrupted speech and brief extracts of speech for example, one word such as ‘yeah’. Asking each participant at the beginning to introduce themselves provided a piece of speech as a basis for identification and upon transcription each participant was given a pseudonym so that each speaker could be identified.

Further familiarisation involved reading and re-reading transcripts and notes recorded during data collection. Notes, thoughts or impressions were recorded whilst doing this; facilitating a developing awareness and thoughts about key ideas or issues. Following familiarisation with the data the researcher was aware of interesting data that may form codes and had started to think about meanings in the data.

#### *4.5.2.3. Open coding of the focus group transcripts*

Conducting open coding of the data ensured that all instances of interesting, relevant data were captured which may not necessarily have been identified by the a priori codes and categories developed from the SRM. This part of the process required logical and intuitive thinking where judgements are made about meaning, importance and relevance of issues and connections between ideas (Ritchie & Spencer 1994). At this stage of analysis, the researcher and one of her supervisor’s each coded a transcript separately and then discussed their findings. As a learning and development exercise this allowed the researcher to share their thinking and check that they could justify and explain their approach to how they coded the data. Although coding is subjective, even if the supervisor and researcher did not come up with the same codes it provided an opportunity for the researcher to consider a different perspective and identify any bias in their coding.

In comparing the coding, the researcher and supervisor had picked up on the same issues although some codes were named slightly differently. Reading the data closely to identify interesting meaningful stories related to PA in HD facilitated coding of

interesting data within the context of the research questions. In terms of eliminating bias during analysis, awareness of one's biases during data analysis reduced the likelihood that data was selectively screened out or included that was only of interest from a certain point of view. For example, although the researcher has a positive view of exercise, this is not to say that people will not have had negative experiences which would have become apparent during the data analysis. In the population studied, it was reasonable that people might have had negative experiences of exercise because of physical limitations or awkward social interactions because of the visual symptoms of HD. The researcher remained conscious that the transcripts should be read with objectivity and all possible data that could contribute to the formation of codes across the dataset in the first read through of the transcripts were included, to then be further scrutinised in subsequent steps of the analysis process.

Following the double coding exercise, the researcher carried out open coding by reading the FG transcripts line by line and assigning 'labels' that summarised or described chunks of what seemed interesting or important data. The researcher carefully read the transcripts and applied a paraphrase or label ('code') that described what they found. The codes were based on the meanings or descriptions that emerged from the data itself. Anything that appeared to be relevant to the research questions from as many perspectives as possible was coded (Gale et al. 2013). The process of coding has also been described as 'linking' because it leads you "from the data to the idea and from the idea to all data pertaining to that idea" (Richards & Morse 2007, p137).

One of the characteristic features of open coding is that of numerous and varied codes being generated (Lofland et al. 2006) demonstrating a thoroughness in the approach. Initial open coding of all of the FG transcripts resulted in the creation of 54 codes (appendix 19). Initial codes with some examples of quotes that they linked to can be seen in table 11.

Table 11: Examples of some initial codes from open coding and quotes linked to them

Quote	Code
<i>"The social side, I suppose. Erm, yes, being able to talk to people, y'know having a little chat to people...I think that's important as well".</i>	What would help them exercise - reasons for exercising
<i>"...he was an excellent swimmer and when we go on holiday he likes to go into a swimming pool but he cannot coordinate himself to swim so that is a big thing for him."</i>	Carers perceptions of limitations caused by HD
<i>"...before when she wasn't doing as much exercise her movements were greater but now her core is a lot stronger she's a lot more in control. She used to...bumping into frames and whether she's able to correct herself quicker I don't know but it's making her a lot more stable."</i>	Perceived positives of exercising or PA
<i>"...the anxiety of going out on their own and people thinking that they're drunk so that just limits people then when their partners come out with them and they're having to hold on to them"</i>	Awareness of HD
<i>"unless you've got a social worker involved and an assessment going on, you're not going to necessarily know that can happen because we didn't know about that before hand."</i>	Support / looking to health and care professionals for answers
<i>"For a carer it's about having a situation where the person you're caring for doesn't have independence which robs me of mine."</i>	Carer's perceptions - burden or difficulties as a carer

The initial codes identified were reviewed to refine them and identify 'codes' for the analytical framework. The researcher referred back to all of the codes one by one to familiarise themselves with the data that had prompted the creation of the code. Firstly, it was noted that there was duplication of one of the codes which was 'hobbies', and so this reduced the number of codes by one. The researcher then continued to review each code and go back to the data it was assigned to.

The code 'co-morbidities' was removed as the discussion was not framed within the context of PA, and the code had not been applied anywhere else, although what was discussed could have implications for PA for this person. Considering this, a note was made to come back to this data later on and see whether it was pertinent to any other codes:

*"goes to a nursing home twice a week for day care and on a Monday she gets weighed....and I keep an eye on her weight ahh... she's been pretty stable for last couple of 12 months but the only thing now, we've got that I didn't know about was (low) blood pressure, I've got to keep an eye on that."*

Similarity of the codes was then looked for and similar codes that represented the same concepts or issues were then grouped together (figure 8). Other examples of analytical coding frameworks have seen up to fifty-nine codes and were also grouped in such a way (Spencer and Ritchie, 1994). Once the codes were grouped together it was easy to look across them and look for repetition in terms of what the names of

codes and what they represented. A number of codes were identified that were essentially the same in terms of meaning but had slightly different names. These codes were collapsed. For example, the codes '*what people with HD and their carer's need to participate in PA*' and '*what carers feel they need to be able to support the person with HD*' were merged. '*What carers feel they need to be able to support the person with HD*' was deleted and '*What people with HD and their carer's need to participate in and support physical activity*' was used as the new name for the code. Going through this process enabled the researcher to refine the codes and reduce the number of codes to be added to the analytical framework, without losing any important aspects identified through open coding. The final refined codes can be seen in table 3.

Categories were developed from the final refined codes (described in the following section). This approach is suggested by Spencer and Ritchie (1994) in their description of the method that the researcher followed, where codes may be grouped and are clearly defined. Having a smaller amount of broad categories to consider which were composed of a number of more specific codes meant that the researcher could go through the transcripts and if something of interest about for example 'relationships' was flagged up, the researcher could then look at the codes of relationship and see which aspect of relationships (i.e. which code grouped under 'relationship') the data related to.

Table 3: Refined codes and final refined codes of the analytical framework identified through open coding (\*merged to form refined code of ‘support’)

Refined codes	Final refined codes
Motivation	Motivation
Environment	Environment
*Support	Support
*What people with HD and their carer's need to participate in PA	
Perceptions of physical activity	Perceptions of physical activity
Reasons for exercising	Reasons for exercising
Caregiver perceptions	Caregiver perceptions
Interactions while exercising	Interactions while exercising
Relationship between caregiver and person with HD	Relationship between caregiver and person with HD
Communication	Communication
Social interactions	Social interactions
Public stigma of HD	Public stigma of HD
Family relationships (stigma)	Family relationships (stigma)

#### 4.5.2.4. Development of categories from the codes

The categories for the analytical framework were developed by grouping the refined codes. The codes were reviewed and those which described similar issues or concepts or appeared to be linked somehow were grouped together with an overarching description that was relevant to them all. These became the ‘categories’ of the working analytical framework formed from the open coding. For example, to the researcher’s thinking, the codes of ‘motivation’, ‘environment’ and ‘strategies’ all described ‘influencing factors’ on PA for the people with HD. Therefore, the terms ‘environment’, ‘motivation’ and ‘strategies’ became codes under the category ‘influencing factors’ in the developing analytical framework. This is illustrated in figure 8 where the process of going from coding, to refining codes and identifying categories is highlighted from left to right. The categories that were developed and their relevant codes can be seen in figure 9.

Figure 8: Example showing how one of the categories of the analytical framework was developed from open coding.

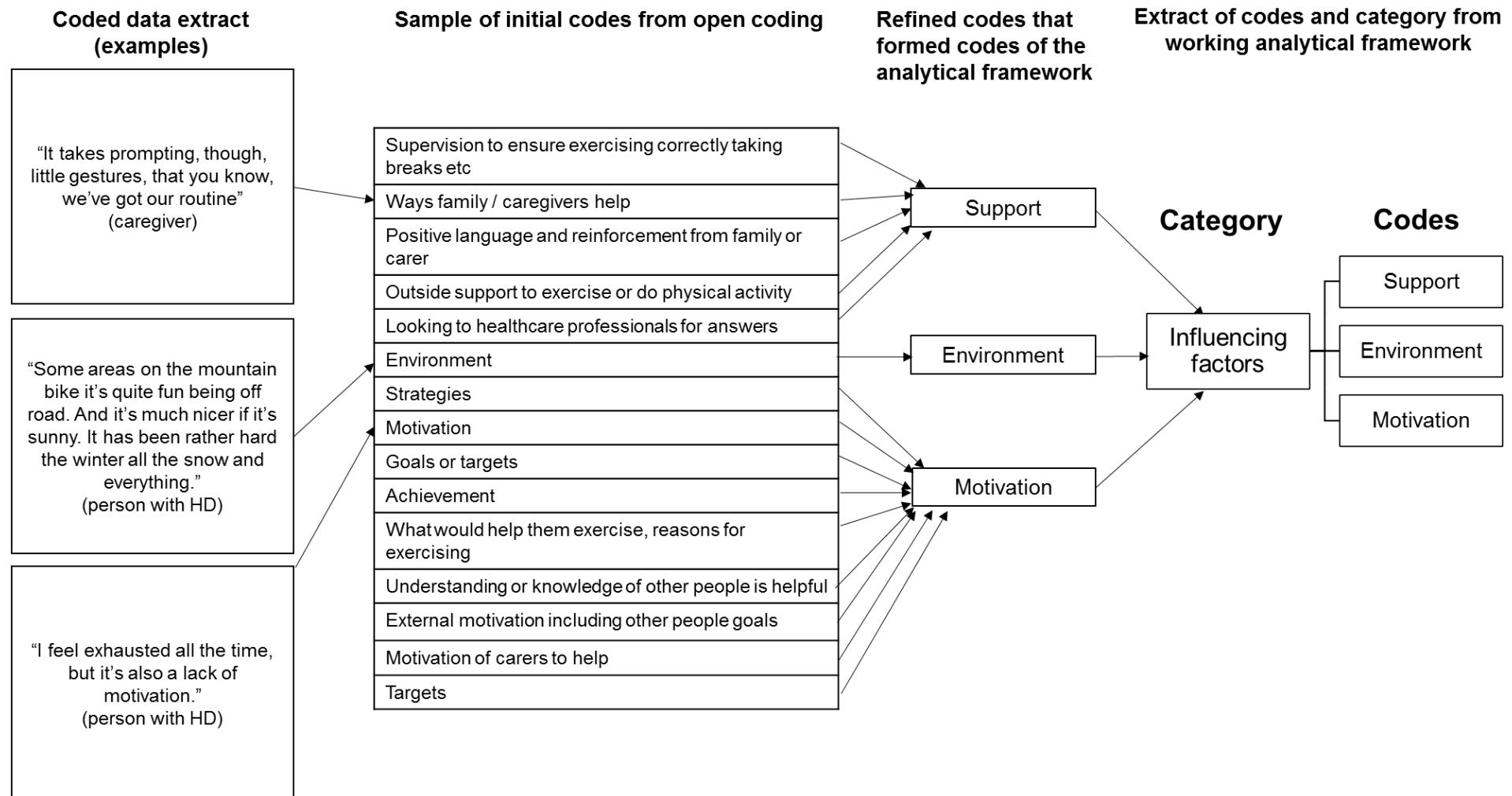
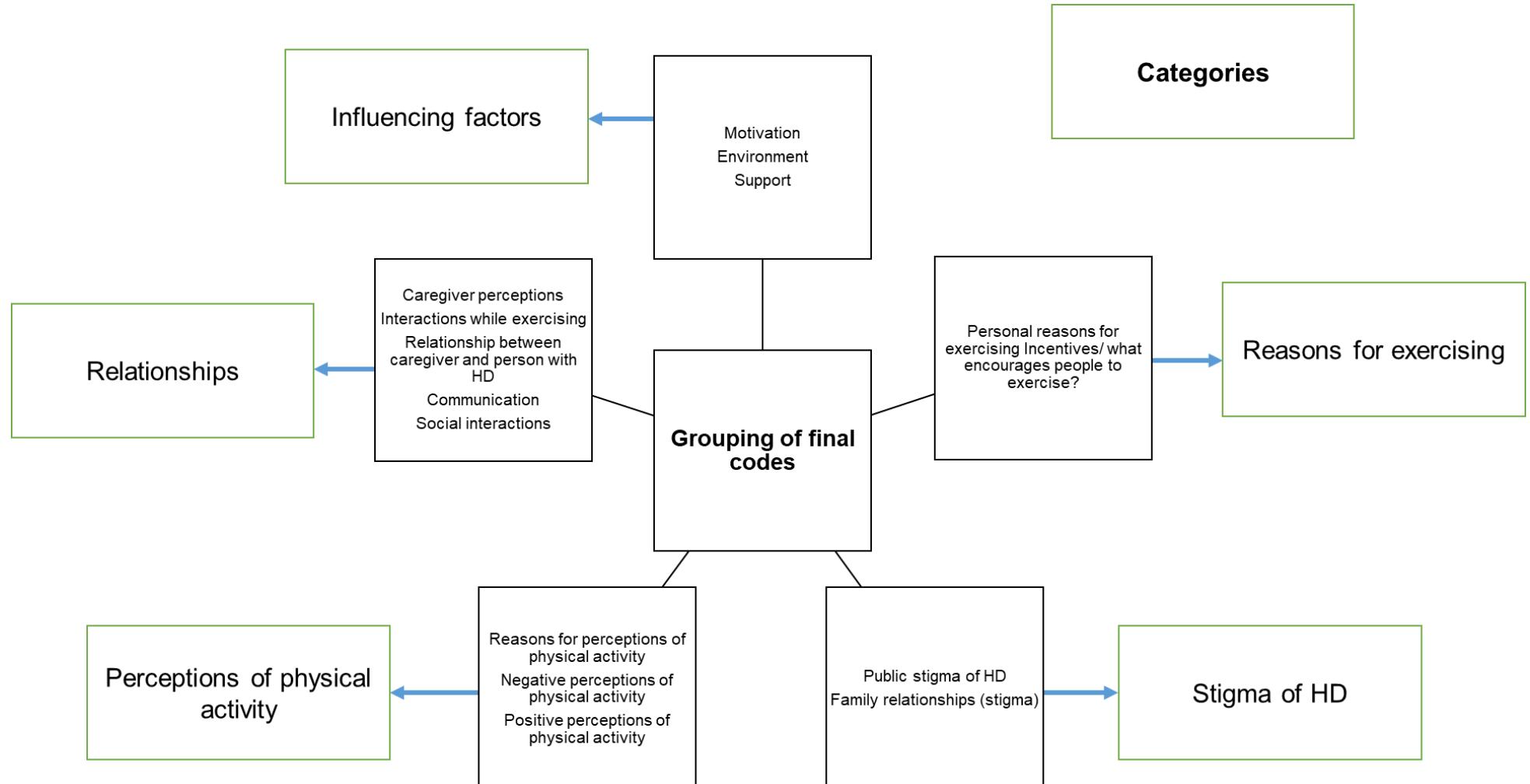


Figure 9: Grouping of final codes to form the categories of the analytical framework from open coding



Descriptions for the codes of the framework were developed and derived from the data so that when applying the coding framework to index the data it was clear to see which data would fit under the code. The descriptions for the codes developed from the data itself were created using the memos the researcher had written and derived from thoughts about the data when creating the codes; an example of a memo regarding identity can be seen in appendix 21.

Figure 9 serves to illustrate how the final codes were grouped and categories were formed, and it essentially illustrates the codes and categories of the working analytical framework derived from open coding. However, the complete working analytical framework including a priori codes and categories, and those developed from open coding is presented in a table format. This is because it allowed space for descriptions of the codes to be included which served as a helpful reminder when indexing the data, and it also made it easy to see the analytical framework as a whole (table 4). The following section discusses refinement of the working analytical framework to produce the final analytical framework to be used.

Table 4: Working analytical framework

Category	Code	Code description
Influencing Factors	Motivation	How motivation (or lack of) influences their physical activity behaviour / attitude towards physical activity.
	Environment	The influence of the environment on physical activity e.g. social / physical environment.
	Support	Different types of support discussed and how that influences physical activity behaviour.
Perceptions of physical activity	Reasons for perceptions of physical activity	Positive or negative – all perceptions of physical activity and possible reasons for those – how they have come to have those perceptions.
	Negative perceptions of physical activity	
	Positive perceptions of physical activity	
Reasons for exercising	Personal reasons for exercising	Why people with HD exercise – their reasons/motivations.
	Incentives/ what encourages people to exercise?	
Relationships	Caregiver perceptions	Impact of caregiver perception on the physical activity of people with HD.
	Interactions while exercising	Interactions whilst exercising - with others / exercise trainer / family member or caregiver.
	Relationship between caregiver and person with HD	The relationship between caregiver and person with HD; how this may impact on physical activity.
	Communication	Communication between people with HD and others – influence of this on physical activity.
	Social interactions	Social interactions in the context of physical activity
Stigma of HD	Public	Other people's lack of understanding or knowledge of the disease and consequences of this for physical activity.
	Family relationships	Stigma/breakdown/difficulties in family relationships because of HD
Coping	Strategies: physical activity	Specific strategies used to overcome limitations or barriers in relation to physical activity
	General strategies	General strategies of coping used
	Normality	What is normal to participants at different stages in terms of physical activity?
	Adapting	Adapting behaviour and/or approach to physical activity because of the limitations caused by HD.
Evaluation	Reflection	Reflecting on specific experiences related to physical activity.
	Emotional experiences	Emotional responses to experiences – how do people react emotionally to certain experiences, what or how does this impact what they do in terms of PA?
	Objective response	How have people responded in a practical way to an experience they have reflected on – i.e. do they respond by troubleshooting, adapting what they do, how they do PA, ask or help or advice?
Cause	Lifestyle	Health behaviours - behaviours such as PA influencing health – what are people's perceptions of this, how does this influence their behaviour.
	Inheritance	Inheritance of the disease – chance of passing it onto children – is PA considered in terms of health behaviour to maintain health?
Consequences	Impact of symptoms	Impact of HD on physical, social, psychological function.
	Continuity	Perspective of how life will continue (also in terms of PA) in the context of the consequences of HD.
	Quality of life	Perspectives with regard to quality of life, how is it altered? Does PA mean anything in terms of improving or maintaining quality of life?
Control	Managing	Perceptions of treatment / management of HD.
	Healthcare	Contact with healthcare professionals.
	Health behaviour	Any perspectives of or beliefs in preventative health behaviours including PA.
	Control over PA	Meaning or perspectives associated with control when it comes to PA related behaviour.
Identity	Impressions of HD	How their impressions/ perceptions of the disease shape their attitudes/perceptions towards PA.
	Living with HD	How experiences of living with the disease shape their experiences and how they relate to physical activity
	Symptoms	How signs and symptoms of HD shape experiences of PA, meaning of the symptoms in the context of PA.
	Family	As a genetic disease how family members deal with HD family history.
Timeline	Change over time	Anticipated change over time and progression of the disease.
Integration of perceptions with memory or experiences	Perceptions affected by experiences	Integration of perceptions with memory or experiences.

#### *4.5.2.5. Testing and refinement of the analytical coding framework*

The ‘working’ analytical framework was tested by two other individuals (visiting fellows to the University of the European Huntington’s Disease Network who work with people with HD). There were numerous benefits to this. Firstly, it allowed the researcher to make sure that they themselves were clear about how the framework should be used because they had to explain it to those who were going to test it. It also gave the opportunity to check that somebody other than the researcher could understand and use the analytical framework. Finally, it afforded the opportunity for discussion around the framework and therefore an opportunity to refine it (if needed) and finalise the framework ready for the next stage of analysis.

The fellows were given transcripts from the focus groups to apply the framework to. They were asked to familiarise themselves with the transcripts and with the codes and categories of the analytical framework (they were given hard copies of the framework, as in table 4). Following this, the fellows were asked to make notes on the transcripts wherever they saw something relevant to any of the codes and put the name of the code next to the data extract (a sample can be seen in appendix 18). As they were familiar with both the transcript and the analytical framework they were able to identify anything interesting that did not seem to have been considered within the framework and should be added.

At the end of the exercise the transcripts and analytical framework were reviewed by the researcher and the two fellows together. There was agreement between the researcher and fellows in terms of what was identified in the transcripts from applying the analytical framework. Of note, the idea of change over time was identified as a potentially important thread within the data and generated interesting discussions which allowed the researcher to explore thoughts and share ideas about the data. Discussion facilitated by this exercise helped to confirm that the analytical framework was usable and understandable. As a result of the exercise, none of the codes or categories were changed, but the description of the code ‘adapting’ within the category of ‘coping’ was altered to also encompass the idea of ‘accepting or not accepting change’ in the context of PA in HD. The final analytical framework can be seen in table 5.

Table 5: The final analytical framework

Category	Code	Code description
Influencing Factors	Motivation	How motivation (or lack of) influences their physical activity behaviour / attitude towards physical activity.
	Environment	The influence of the environment on physical activity e.g. social / physical environment.
	Support	Different types of support discussed and how that influences physical activity behaviour.
Perceptions of physical activity	Reasons for perceptions of physical activity	Positive or negative – all perceptions of physical activity and possible reasons for those – how they have come to have those perceptions.
	Negative perceptions of physical activity	
	Positive perceptions of physical activity	
Reasons for exercising	Personal reasons for exercising	Why people with HD exercise – their reasons/motivations.
	Incentives/ what encourages people to exercise?	
Relationships	Caregiver/family perceptions	Impact of caregiver perception on the physical activity of people with HD.
	Interactions whilst exercising - with others / exercise trainer / family member or caregiver	Interactions whilst exercising - with others / exercise trainer / family member or caregiver.
	Relationship between caregiver/family member and person with HD	The relationship between caregiver and person with HD; how this may impact on physical activity.
	Communication	Communication between people with HD and others – influence of this on physical activity.
	Social interactions	Social interactions in the context of physical activity
Stigma of HD	Public	Other people's lack of understanding or knowledge of the disease and consequences of this for physical activity.
	Family relationships	Stigma/breakdown/difficulties in family relationships because of HD
Coping	Strategies: physical activity	Specific strategies used to overcome limitations or barriers in relation to physical activity
	General strategies	General strategies of coping used
	Normality	What is normal to participants at different stages in terms of physical activity?
	Adapting	Adapting behaviour and or approach to physical activity because of the limitations caused by HD, accepting or not accepting change in relation to physical activity in HD
Evaluation	Reflection	Reflecting on specific experiences related to physical activity.
	Emotional experiences	Emotional responses to experiences – how do people react emotionally to certain experiences, what or how does this impact what they do in terms of PA?
	Objective response	How have people responded in a practical way to an experience they have reflected on – i.e. do they respond by troubleshooting, adapting what they do, how they do PA, ask or help or advice?
Cause	Lifestyle	Health behaviours - behaviours such as PA influencing health – what are people's perceptions of this, how does this influence their behaviour.
	Inheritance	Inheritance of the disease – chance of passing it onto children – is PA considered in terms of health behaviour to maintain health?
Consequences	Impact of symptoms	Impact of HD on physical, social, psychological function.
	Continuity	Perspective of how life will continue (also in terms of PA) in the context of the consequences of HD.
	Quality of life	Perspectives with regard to quality of life, how is it altered? Does PA mean anything in terms of improving or maintaining quality of life?
Control	Managing	Perceptions of treatment / management of HD.
	Healthcare	Contact with healthcare professionals.
	Health behaviour	Any perspectives of or beliefs in preventative health behaviours including PA.
	Control over PA	Meaning or perspectives associated with control when it comes to PA related behaviour.
Identity	Impressions of HD	How their impressions/ perceptions of the disease shape their attitudes/perceptions towards PA.
	Living with HD	How experiences of living with the disease shape their experiences and how they relate to physical activity
	Symptoms	How signs and symptoms of HD shape experiences of PA, meaning of the symptoms in the context of PA.
	Family	As a genetic disease how family members deal with HD family history.
Timeline	Change over time	Anticipated change over time and progression of the disease.
Integration of perceptions with memory or experiences	Perceptions affected by experiences	Integration of perceptions with memory or experiences.

#### *4.5.2.6. Indexing of the transcripts*

Once finalising of the analytical framework was complete, it was used to index the data within the transcripts. NVivo 9.0 was used to index the transcripts. Computer software has previously been used to organise and sort qualitative data following the framework method (Ward et al. 2013). At this stage it was easier to store, sort, and organise the data in this way due to the volume of paper there would have been if doing the indexing by hand.

The analytical framework was created in NVivo by adding categories as 'parent nodes' and adding codes of the framework as 'child nodes' (i.e. sub sections of the nodes) (appendix 17). The transcripts were imported into NVivo 9.0 and the researcher reviewed each transcript, indexing the data to relevant nodes. NVivo 9.0 facilitated a systematic approach to indexing the data which was achieved by 'highlighting' selected text within each of the transcripts and ascribing it to the relevant code/category of the framework (i.e. the relevant 'parent' or 'child node' representing the code of the framework created within NVivo 9.0). The sections of the text were 'tagged' to the relevant code of the coding framework by highlighting it, right clicking, selecting 'code selection' and selecting the relevant 'node' (code) from the list. On completion, all of the data was indexed to the relevant 'nodes' (code). Descriptions that had been developed for the categories of the analytical framework helped the researcher to ensure that only relevant data was indexed to the analytical framework codes and categories.

Interesting stories, comments and dialogue between participants were indexed that captured individual responses and group responses (which would also allow comparison across groups at the mapping stage of the analysis). Not every single line was indexed because of the tendency for focus groups to 'go off subject' every now and then on an unrelated tangent. One must also be cognisant that the group dynamics and interaction would have influenced the discussion within the group and as a result, perspectives would have arisen and possibly been modified (May 2000). Reflecting on these issues led the researcher to utilise an approach that focussed on analysing both what the individuals actually speak about, and the stories that form the focus for the group (Kidd and Parshall 2000). Such flexibility allowed for emergence in analysis of any overall agreements in the groups (and indeed to compare across the groups) but also to identify where any sense of group consensus could in fact be artefact by comparing it to the responses of the individuals. All relevant participant discussion including whether any individuals particularly influenced the group was noted (May 2000). This was achieved by closely reading line by line, which is suggested to 'alert

the researcher to consider that which may ordinarily remain invisible because it is not clearly expressed or does not ‘fit’ with the rest of the account’ (Braun and Clark 2006). This is thought to challenge the developing analysis to explain anomalies or deviant cases which also makes the analysis stronger. Some examples of data that was indexed to the analytical framework can be seen in table 6 overleaf. While indexing the data, the researcher used the ‘annotations’ function in NVivo 9.0 to make any notes of thoughts and ideas which may help to consolidate the data at later stages when forming the key themes. An example of this can be seen in appendix 20.

Table 6: Indexed quotes and relevant codes/categories of the analytical framework

Indexed quote	Code / category
<i>"I used to do all those things a real keen sportsman; football, cricket but this illness means that I'm not steady on my feet."</i>	Change over time / Timeline
<i>"Gardening... I was just generally walking you know... out across the moor when I could...as I said that's all finished. Finished now"</i>	
<i>"It's a good feeling so yeh it's like um a good feeling at the moment I'll try and keep that up for as long as I can...I worry that everything takes me longer, I look at the clock and think "oh, is it that time already" because everything takes longer when you get Huntington's."</i>	
<i>"I do attempt a lot more exercise (since diagnosis of HD), I just happen to be, I have this motivation but erm... motivation is to go out and get fresh air. I live on my own so, erm, my children aren't as in touch as they used to be. I know it is my responsibility."</i>	
<i>"I trip a bit, you know, I sort of....hum, I'm not good on my feet...hum going down stairs I go down backwards, but I've been like that for a long time a couple of years, you know"</i>	Impact of symptoms / Consequences
<i>"...like I said XXX's brother and sister's got it, her brother has passed away now and the all three have got something different, there's not one thing in common, well there are one or two things that are common to them all like the involuntary movement and all that, but speech is different for some, the mind is effected more with others, so each individual have got so many different symptoms that is not one common thing"</i>	
<i>"...but you know, that's been sorted but to get her motivated and actually to get her out, she was quite happy just to be sat just looking at the same four walls and to me, she was just surviving, she wasn't living. And I think that's what started it (encouraging the woman with HD to start building a physical activity routine)..."</i>	Quality of life / Consequences

#### *4.5.2.7. Charting data into the framework matrix*

Spencer and Ritchie (1994) discuss charting the data by lifting it from the original textual context (the transcript) and placing it into charts or matrices consisting of headings and sub-headings which are actually the names of the codes and categories that comprise the analytical framework. When text within the transcripts are indexed to nodes, NVivo 9.0 also automatically places that data within the relevant 'node', i.e. it also lifts the data from the original transcript and places it into the 'node'. In essence, it begins the process of charting where the data indexed to each node can be seen within that node by double clicking on it (a new window opens containing the node name and all data indexed to it).

At the point where indexing was complete and it was possible to look within each node and see all of the data indexed to it, the researcher created charts/matrices within Microsoft Word using tables. The categories and codes of the analytical framework were used to create the headings of the matrix for the data in a table and the data was lifted (copied) from each 'node' in NVivo 9.0 and pasted under the relevant heading (an example of can be seen in table 7), i.e. moved across into the chart manually by the researcher.

Table 7: Example of category in analytical framework matrix

FOCUS GROUP NO.	CATEGORY: <b>INFLUENCING FACTORS</b>
	Code: <b>Environment</b>
1	P11: Like going on holiday and having to walk a bit further than you usually do I think that would be quite helpful and I'm going to do that in a week's time

Due to the volume of data, it was particularly challenging to look across the data and so at this point, the charts were printed out. Once all data was charted in this way and the numerous sheets printed out and laid out, it was possible to start looking across the codes and categories to begin forming descriptive themes. The way that the data was presented in the framework matrix also made it possible to look within focus groups (FGs) at individual cases and group consensus and across FGs in terms of the data that had been coded to each code of the framework. The aim was to refine and develop the codes and categories into 'themes'; key themes and sub-themes. Having the data organised in such a way facilitated the next stage of mapping and synthesising the data.

#### *4.5.2.8. Mapping and synthesis of the data to create key themes and sub themes*

Once all of the data was charted into the framework matrix it was possible to look across all of the codes and categories of the framework and see how they were represented across the dataset. With the data organised in this way the researcher was able to start identifying initial and eventually key themes. This was achieved in a number of ways. These included identifying emergent patterns and commonalities across the charted data and checking whether the date actually did fit into the codes it had been indexed to. The researcher also looked for overlap and repetition across codes and categories, and thought about whether there were better descriptions that encompassed data from more than one code that could describe the data and thus form an initial theme (Ryan and Bernard 2003).

Firstly, data within the codes were scrutinised for similarities and overlap of key issues. The purpose of this was so that codes (and even categories if appropriate), could be collapsed into each other where the data justified the formation of an overarching theme. Two categories that were collapsed into one were ‘integration of perceptions with memory or experiences’ and ‘evaluation’. ‘Integration of perceptions with memory or experiences’ had quotes linked to it that seemed more relevant to ‘evaluation’. Therefore, data from the two categories were collapsed into the code ‘evaluation’ and the code ‘integration of perceptions with memory or experiences’ was deleted. At this point, ‘evaluation’ was noted as an ‘initial theme’. Following further scrutiny of the data, this initial theme was merged with other initial themes identified as ‘adapting’ and ‘coping’ as the reflections and evaluations of PA experiences linked to how people had adapted or responded to HD symptoms and progression. Data within the framework categories of ‘control’ and ‘cause’ were also considered relevant to this developing thread.

Reviewing the data of these codes at this point revealed that an overarching idea of coping in relation to PA in HD was emerging. The concepts of adapting, trying to maintain normality, independence, overcoming limitations, managing change, understanding and accepting limitations in relation to PA were all present. It was challenging to actually consolidate all of those concepts which related to coping into sub themes. There were a number of iterations of the name of the key theme and subthemes that encompassed these ideas before the final one was decided upon (figure 10).

Figure 10: Iterations of the names of one of the key theme and its subthemes

Iterations of key theme and subtheme names				(Final key theme and subthemes)
(Key theme) (subthemes)	1) Coping	2) Coping or non-coping and managing	3) Strategies, struggles and successes	4) Achieving PA participation whilst coping with the nuances of HD
	<p>• Adapting • Normality • Trying to overcome limitations • Coping with or managing change • Understanding or accepting one's limitations</p>	<p>• Adapting • Normality • Trying to overcome limitations • Coping with or managing change • Understanding or accepting one's limitations • Independence • Strategies • Managing • Safety</p>	<p>• Coping / non-coping • Adapting • Managing • Strategies • Control • Independence</p>	<p>• Continually adapting and adjusting expectations of ability over time • Tried and tested: strategies for engagement specific to HD across the stages.</p>

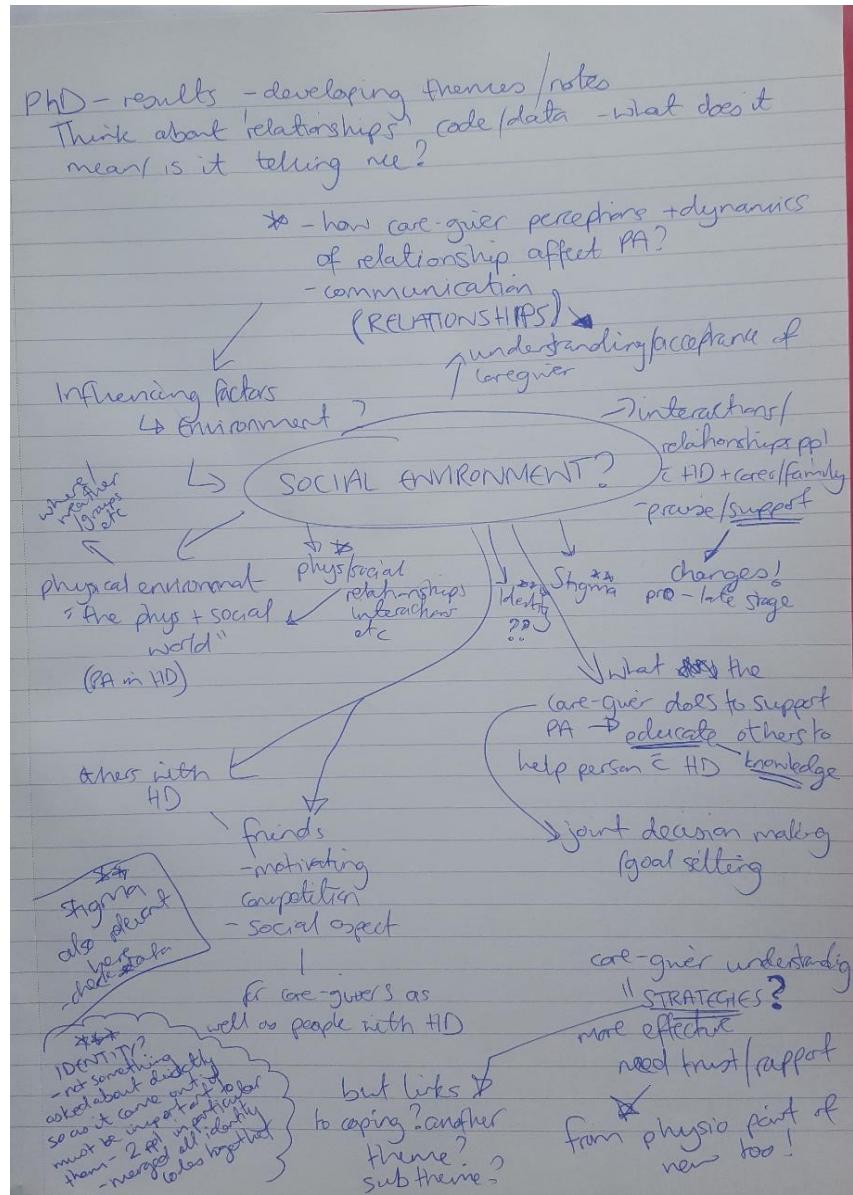
Through a process of comparing, contrasting and making links between the data and codes and categories, initial themes were developed. A key issue for the researcher was ensuring that all of the rich insight the data provided would be represented within the key themes. Developing sub themes within the key themes facilitated this.

Development of another of the key themes started with the researcher reviewing the framework matrices and the data within it. It was noted that the category 'relationships' had a lot of data indexed to it, so the researcher started by scrutinising the data within this category. The codes within the category of 'relationships' were 'caregiver/family member perceptions', 'interactions while exercising', 'relationship between caregiver and person with HD', 'communication' and 'social interactions'. However, when looking across the data that had been indexed to these codes in the matrix, and thinking about what the emerging 'message' seemed to be, it became apparent that the idea of 'social environment' was more pertinent as a dominant overarching concept that encapsulated what the data described (see table 8 for a sample of the data to support this). This thought process was captured in a memo (figure 11).

Table 8: Example of some quotes indexed to codes and categories of the analytical framework that related to the concept of the 'social environment' as an initial theme.

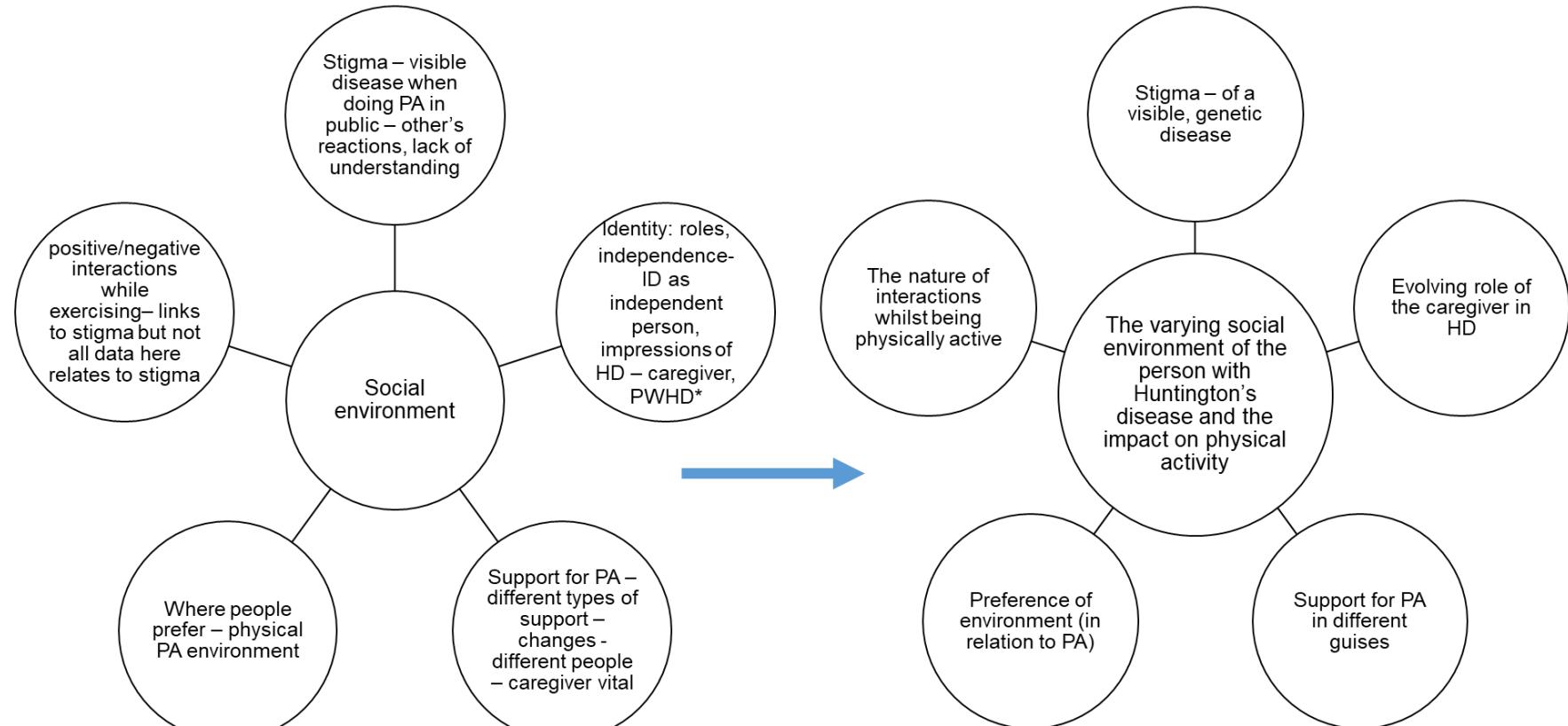
Quote	Code / category in analytical framework	Initial theme
"When he went to the day centre he didn't like it and he wouldn't go, yeah. It wasn't for him is what he felt. And our social worker at the time said 'well, what would he like to do?' and the thing that came up really was the gym. Er, so the fact that they fund this personal trainer now through direct payments has enabled him to do it. I do the transportation. Well now he's embarrassed about going to the gym, erm, she comes to the house and she'll work with him in the garden and do circuits. She'll take him to the beach and do body boarding and different things like that. She shakes it up a bit."	Support / influencing factors	Social environment
"I suddenly realised that it was about 2 hours per day on the bike it's a bit more than an ordinary session of exercise...some areas on the mountain bike it's quite fun being off road. And it's much nicer if it's sunny. It has been rather hard the winter all the snow and everything."	Environment / influencing factors	
"she felt she could walk enough so she could go to the bus stop and catch a bus in, she had her bus pass and she was for a couple of years able to go to the bus stop, get on a bus and go to [place], but then she had a altercation with a bus driver, he thought that she was drunk on drugs or something like that"	Public / stigma	
"...there became a time when he was too embarrassed to go to the gym because he felt people were looking at him. And you know, they were! I had to go and support him in the gym when he was doing his weights because of balance and co-ordination and things."	Public / stigma	
"I think it's good to um be social and be with other people, interacting as well as you can you know? To a point where they stop noticing things."	Interactions when exercising / relationships	
"I was used to doing the, umm, going in and caring for someone. Whereas it's, with [name], it's enabling her to stay independent. Not doing it for her. It's having the time, the patience and the understanding."	Relationship between care-giver and person with HD / relationships	
"Don't see them just as a HD sufferer. They need to be seen as an individual who has HD and HD is XYZ, but this is the person's needs."	Living with HD / identity	
"I'm now doing the gym. So it used to be somewhere I can go and I've also got balance classes there as well which I just started doing as well so 3 times plus a half hour balance as well. But it's taken me a long time to get to that and then if I don't go I feel myself drifting back and it doesn't take long before I start getting pain in my joints again. I have also got support shoes because of an Achilles tendon. I thought that was something to do with Huntington's. But it's not (laughs)! So my funny walk is also different again because it also hurts me unless I've got my right shoes on (laughs). So but, yeah, as long as I seem to have the right shoes then it helps with being able to do the exercise."	Symptoms / identity	
"Sometimes you have to accept it and not push, (that the person with HD does not want to take part in activities) cos that's the nature of her illness, and so you just can't do it for her so but in time she'll make that choice for herself, and when she does no one'll (no one else will) get a go on that bike!"	Impressions of HD / identity	

Figure 11: memo relating to development of key theme related to social environment



Following the identification of 'social environment' as a potential theme, other codes and categories were reviewed to see whether the data was also relevant to social environment. The categories of 'stigma' and 'identity' were also identified within the framework matrix as being of relevance and linked to the 'social environment'. These two categories also encapsulated data that gave insight into the social settings and experiences of PA of the individuals with HD. It was helpful to create spider diagrams to map and refine the potential key theme and sub-themes to visualise how and where they may fit together (figure 12). Once the researcher was satisfied with the composition of the theme, the names given to the sub themes and key theme were reviewed finalise them and to check that the names were a good description of what the data entailed (see researchers notes in figure 12).

Figure 12: Development of a key theme; spider diagrams and memos



1<sup>st</sup> spider diagram - First go at developing social environment as key theme from my notes when looking across the analytical framework matrix. Looking at this and the data, identity as a subtheme may fit better in with the developing theme related to representations of the illness and how that impacts on PA although it is also relevant here. Support for PA- I think that actually the role of the caregiver should stand alone looking back at the data indexed under the category of 'relationships' in the analytical framework matrix because the role of the caregiver is coming across as vital to a lot of participants! Support is not only from the caregivers though so maybe better to split 'support for PA' into 2 subthemes so it's all captured appropriately.

2<sup>nd</sup> spider diagram - From the first spider diagram – have created 2 subthemes from 'support for PA' = Support for PA in different guises' and 'evolving role of caregiver in HD'. Identity has been taken out to be incorporated into key theme 1. I've thought more about the names of the sub themes and key theme to capture the essence of what they describe and highlight in terms of how HD impacts of PA in terms of the social environment.

\*PWHD = person with HD

Data analysis also identified another key theme. In reviewing the data within the matrix, the category of 'perceptions of PA' encompassed data that gave insight into perceived benefits, terminology that people use, motivations to do PA and reasons for engaging in PA. This clearly linked to the category of 'reasons for exercising', but links with other categories were also observed because of the broad data indexed to it, (due to it being a broadly named category). For example, perceptions of PA were informed and affected by the participants' social environment, illness representations, (consequences, timeline, control, identity).

Considering the research questions were focussed around PA, perceptions of PA would have emerged as a major thread, but it is how those perceptions linked in to the nuances of living with HD and how the researcher captured those insights that was important. Although 'perceptions of PA' was considered as a potential stand-alone key theme, the researcher reasoned that exploring how perceptions of PA weaved into and across other key themes that encapsulated different aspects of social environment, coping and illness representations would give a richer insight. Although in the abstract sense, different perceptions of PA are captured across the key themes, in terms of the physical data analysis, the category 'perceptions of PA' was explicitly merged with 'reasons for exercising' because the data overlapped between these two categories. This was renamed to form two subthemes of 'reasons and motivations for PA' and 'expectations and experiences of benefit'. These linked strongly to the category of 'consequences', i.e. from the data, the consequences of HD were seen to impact on reasons for, and experiences and expectations of PA.

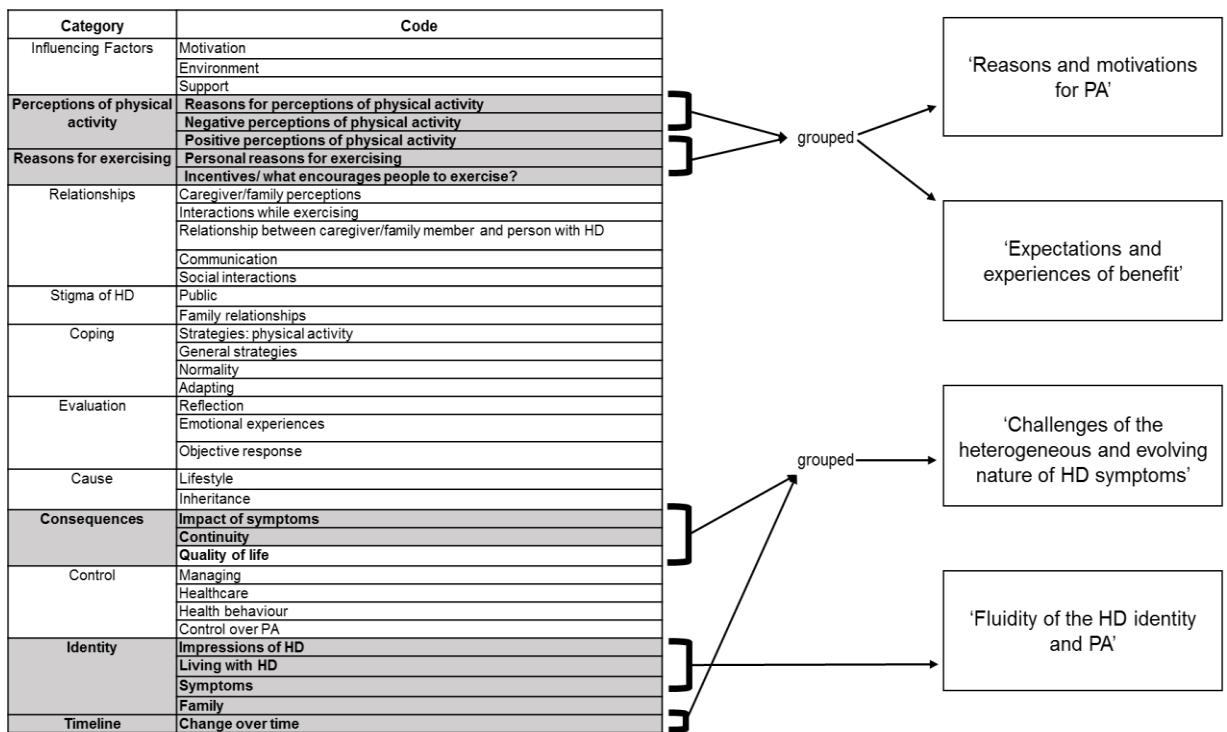
On first review of the data within the category of 'consequences' which encompassed the codes 'impact of symptoms', 'continuity' and 'quality of life', the researcher considered this as a potentially overarching key theme because of the rich insight and volume of data it represented. However, it was clear that change over time was a major thread within the data and so the researcher looked to the category of 'timeline' to see whether the data there could be grouped with 'consequences', and was found to be relevant. Subsequently the categories of 'consequences', and 'timeline' within the analytical framework matrix were grouped together to form the subtheme of 'challenges of the heterogeneous and evolving nature of HD symptoms. Table 9 depicts quotes that relate to the categories of timeline and consequences. Within the quotes indexed to consequences, although they primarily relate to the symptoms of HD, the element of time is present, i.e. 'the longer we can keep it', 'doesn't swim anymore'.

Table 9: Quotes indexed to categories of consequences and timeline

Quote	Code / category in analytical framework
<i>"I think that, you know, possibly in the early stages of HD hum exercise would be a great thing...as you get deeper into the disease you are unable, physically unable, and probably mentally as well hum to exercise. I think with all the goodwill in the world, you know, they'd love to do it, but they are unable as they get further into it."</i>	Change over time / timeline
<i>"it's taken me in the last few years or so to...what with everything that's been going on recently with the diagnosis and all of that I just didn't want to do anything. I was just sitting at home but it took a lot to then get me in to something else afterwards. To start doing the gym...I couldn't do the badminton because I didn't have the co-ordination anymore."</i>	Change over time / timeline
<i>"He [doesn't swim anymore] can't swim in a straight line. Say this is the lane, he'll swim diagonally as he hasn't got the co-ordination..."</i>	Impact of symptoms / consequences
<i>"it's involving some level of physical activity which can constitute as exercise and it seems to function quite well and I think the longer we can kind of keep it in that place the better off were going to be."</i>	Continuity / consequences

Although originally included in another key theme, the category of 'identity' was instead included in this theme. This was because as the theme evolved, it was clear that the concept of 'identity' naturally fitted with the emerging messages of this key theme which relate to the continuously evolving representations of HD that participants experience and how that impacts on PA. Figure 13 illustrates the subthemes of this key theme and how they emerged from the data indexed to the categories and codes of the analytical framework. The name for this key theme was refined from 'illness representations' to 'the evolving representations of Huntington's disease and their impact on physical activity'. This name gives a better representation of what the theme describes in terms of the changes over time, the consequences of HD and the impact on participant perceptions across the stages.

Figure 13: Illustration of how subthemes for this key theme emerged from the data indexed to the categories and codes of the analytical framework.



Through following the process described for all of the data in the matrix, development of key themes and subthemes that mapped the emerging issues in the data and illuminated the experiences of PA for the people with HD was possible.

The key themes are presented and discussed in detail in the following results and discussion chapters.

## 4.6. Ethical considerations

### 4.6.1. Ethical approval

Ethical approval was gained from the School of Healthcare Sciences, Cardiff University to conduct the research (appendix 11). As participants were recruited through the Huntington's Disease Association (HDA), ethical approval for the research was required only from the School of Healthcare Sciences, Cardiff University.

### 4.6.2. Informed consent

A flyer containing information about the FG sessions was circulated to potential participants via the local Huntington's Disease Association specialist advisors (example in appendix 8). Further information was given within information sheets to those who attended the FG sessions (appendix 9). Participants had the opportunity to ask

questions for clarification on any issues regarding the research. Fully informed and freely given written consent was obtained from FG participants before the start of the sessions. The researcher had received up to date training in taking informed consent and good clinical practice.

#### **4.6.3. Data management**

The research was conducted in accordance with the recommendations for physicians involved in research on human participants adopted by the 18th World Medical Assembly, Helsinki 1964 and later revisions. All work undertaken as part of this study complied with the Research Governance Framework for Health and Social Care in Wales and the Cardiff University Research Governance Framework. All participant identification procedures and procedures for data storage, processing and management complied with the Data Protection Act 1998. The Huntington's Disease Association (HDA) maintain a confidential mailing list for members of the HDA who have agreed to be used for postal contact allowing them to approach people about taking part in research studies. The workshop information was circulated to everybody on the care advisors' databases in each region by post. All correspondence was initiated by the HDA and no personally identifiable details were provided to the researcher without the consent of the involved individuals.

#### **4.6.4. Confidentiality and anonymity**

The researcher maintained preservation of confidentiality and anonymity of participants in accordance with the Data Protection Act 1998 (Parliament 1998). All participants were allocated a unique identifier to be referred to in the transcripts and all data collected was held in a linked anonymised form. Audio files were copied onto a password protected computer so that only the researcher had access and were deleted from the Dictaphone. Electronic audio files were kept on a password protected computer whilst being transcribed and destroyed on research completion. To ensure participant anonymity, details including names and locations were omitted from the transcripts and replaced with [place] / [name]. Consent forms were stored in a locked filing cabinet at the School of Healthcare Sciences.

## 5. Results and discussion of key themes

### 5.1 Chapter overview

The following chapter presents the research findings. Firstly, the descriptive demographic data of the focus group (FG) participants is presented. A brief introduction to the key themes is then given. This is followed by presentation and discussion of the three key themes in three consecutive chapter sections. Lastly, the main discussion section draws together the key themes and discusses them in relation to the theoretical model chosen to underpin this research; the Self-regulation model (SRM).

In the presentation of results, quotations from the transcripts have been used to support the findings reported, illuminate experiences described by participants and illustrate certain points. Examples of quotes from the FGs that are used sometimes have the following notation '[...]'. The purpose of this is to shorten quotes so that the essence of what is being said is not lost but makes the quotation more succinct for the reader. Additionally, words that are presented in [brackets] have been included in quotes to remove distinguishing features such as names and specific locations.

### 5.2. The Focus groups

Eight focus groups (FGs) were conducted across the UK. The questionnaire data collected from participants (described in the methodology chapter) were used to give context as to the 'make-up' of the FGs i.e. 'description' in the demographic data tables (appendix 16). In the following section a description of each FG is given in terms of the FG participants and dynamics (see table 10 for details of numbers and make-up of the groups in terms of people with HD, caregivers and professionals).

The FGs consisted of a mix of people with HD (or gene positive), caregivers who were family members or in employment and the specialist HD advisors who had organised the session in their location. There was also one exception where in the one of the groups, a physiotherapist and physiotherapy assistant attended. This was because they worked at an assisted living facility and had been approached by the local HDA specialist advisor and so brought residents with them as they were also interested in discussing physical activity (PA) in HD. The physiotherapist and physiotherapy assistant knew the residents well and worked closely with them on a daily basis. Some were late stage and had speech impairment and needed their support to attend the FG. The various FG locations allowed the researcher to access people of diverse

backgrounds but acknowledges that the findings are not exhaustive of how living with HD impacts PA participation. Participants with HD covered the spectrum of the disease from prodromal to early, mid and late stages. FG2, FG4 and FG8 consisted of more people at early stage HD (see table 10). FG3 consisted of people with HD who were at mid-late stage. Participants of FG7 with HD were early to mid-stage. FG1, FG5 and FG6 included a range of people from early to mid-late stage HD. Amongst the FG discussions there were no major disagreements so much as alternative perspectives and varied experiences that shaped those perspectives. Participants remarked that they found the conversations and sharing of experiences useful for themselves for what they could do in terms of PA. In some cases, participants (caregivers in particular) remarked that the sessions were a positive experience for them and the people with HD in that they were 'given the floor' so to speak, were listened to and experiences and advice were shared. It is likely that the structure of the FG sessions contributed to fostering a relaxed environment because people were given the chance to meet each other and circulate before the sessions whilst having refreshments.

FG1 participants were a mix of people with HD, caregivers (family members) and professionals. Generally, there was a good contribution from different group members. There was a clear understanding within the group that the people with HD needed longer to speak. Those with HD spoke more quietly than the rest of the group but were encouraged to finish what they wanted to say rather than being spoken over or spoken for. Most people knew each other and were aware of the speech impairments and cognitive difficulties that mean for some, they need longer to process the questions and respond. A unanimously held view amongst all participants was that PA of some description was an important 'intervention' for HD specifically.

FG2 included professionals, people with HD and caregivers. The dynamics within this group fostered an environment where discussion and sharing of experiences sometimes turned to general advice giving (by the male caregiver to the one male with HD). The researcher emphasised to the person with HD that the Specialist HD advisor was there to listen and offer advice regarding any concerns following the FG session. The flow of conversation went in the directions of sharing of concerns and wanting to problem solve ideas about making PA more accessible for people with HD. One particularly vocal participant (male caregiver) might have influenced some of the discussion with his views, but not necessarily alter other participants' views.

FG3 was in the same location as FG2. It was clear that a rapport between the participants developed quickly. Sharing advice sharing, experiences and empathising

recurred throughout this FG session. FG4 was one of the smaller FGs and as such, the participants each shared rich detail of their experiences and perspectives about HD and PA. The participants with HD were at early stage and were both trying to be physically active. The general perspective that emerged from the FG was that it is good to be physically active.

There was a positive dynamic between participants of FG5 who all knew each other. A supportive environment was created by the participants as they prompted each other to share their thoughts and experiences. At times the physiotherapist or caregiver would ask the participants with HD if it was acceptable to them that they spoke about them in relation to PA; to which the participants agreed and would add to what the person was saying about them or nod if they were in agreement. The participants of FG6 led the discussion towards talking about ways of keeping active other than high level exercise, perhaps reflective of the mid to late-stages that the people with HD were at. Slightly more facilitation was required with this FG as 2 participants had slight speech impairment and they needed to be drawn into the conversation along with the other participants.

Generally, participants of FG7 were physically active in a variety of ways. The caregivers would sometimes help the participants with HD to articulate what they were saying. Amongst this group there were also examples of participants influencing what others might do to become more active through information sharing regarding PA. The dynamics of FG8 changed throughout the discussion. There were numerous points where participants related to other's experience of exercise specifically regarding HD. There was also discussion where advice was given, and participants appeared to inspire each other to be active.

Prior to the FGs, the researcher considered the possibility that most people who attended would be early stage and not many at late stage. This is because of increased difficulties and pronounced apathy that people experience at later disease stages which may make them less inclined to invest their time in taking part in activities not part of their daily routine. When the time came, it appeared that people who were interested in talking about PA and exercise attended regardless of disease stage and so a spread of people across the stages attended.

Table 10: Focus group participants and locations

Focus Group	Site	Participants
1	Suburb; affluent area of mid- England.	<b>13 participants</b> Four people with HD (n=1 female; n=3 males; 2 mid-late stage, 1 early stage) Seven caregivers/family members (n=5 females; n= 2 males 2 specialist HD advisors (n=1 male)
2	City in South Wales	<b>Six participants</b> Two people with HD (n=1 male early stage; n=1 female mid-late stage. Three caregiver/family members (n=2 females; n=1 sister, n=1 spouse; n= 1 male (spouse)) One specialist HD advisor (female)
3	City in South Wales	<b>Five participants</b> Two people with HD (n=2 female's mid-late stage). Three formal caregivers: (n=3 females)
4	Urban city, northern England	<b>Four participants</b> Two people with HD (n=1 female; n=1 male; both early stage) One caregiver/family member (male spouse) One specialist HD advisor (female)
5	Urban city, northern England	<b>Eight participants</b> Four people with HD (n=1 female early stage; n=3 males early to late stage) Two caregivers: (n=1 family member (mother); n=1 female healthcare assistant. Two professionals (n=1 male physiotherapist; n=1 male physiotherapy assistant)
6	Rural location, Southern England	<b>Five participants</b> Three people with HD (n=2 males; mid stage; n=1 female; mid-late stage) Two caregiver/ family members (n=1 male; spouse; n=1 female; spouse)
7	Rural, affluent area of England.	<b>Eight participants</b> Four people with HD (n=1 female; n=3 males) Four care-givers (n=3 females; spouses; n=1 male; spouse)
8	Rural, affluent area of England.	<b>Eight participants</b> Four people with HD (n=4 males; 1 male gene positive) Four caregivers/family members (n=4 females; spouses)

### **5.3. Introduction to the key themes**

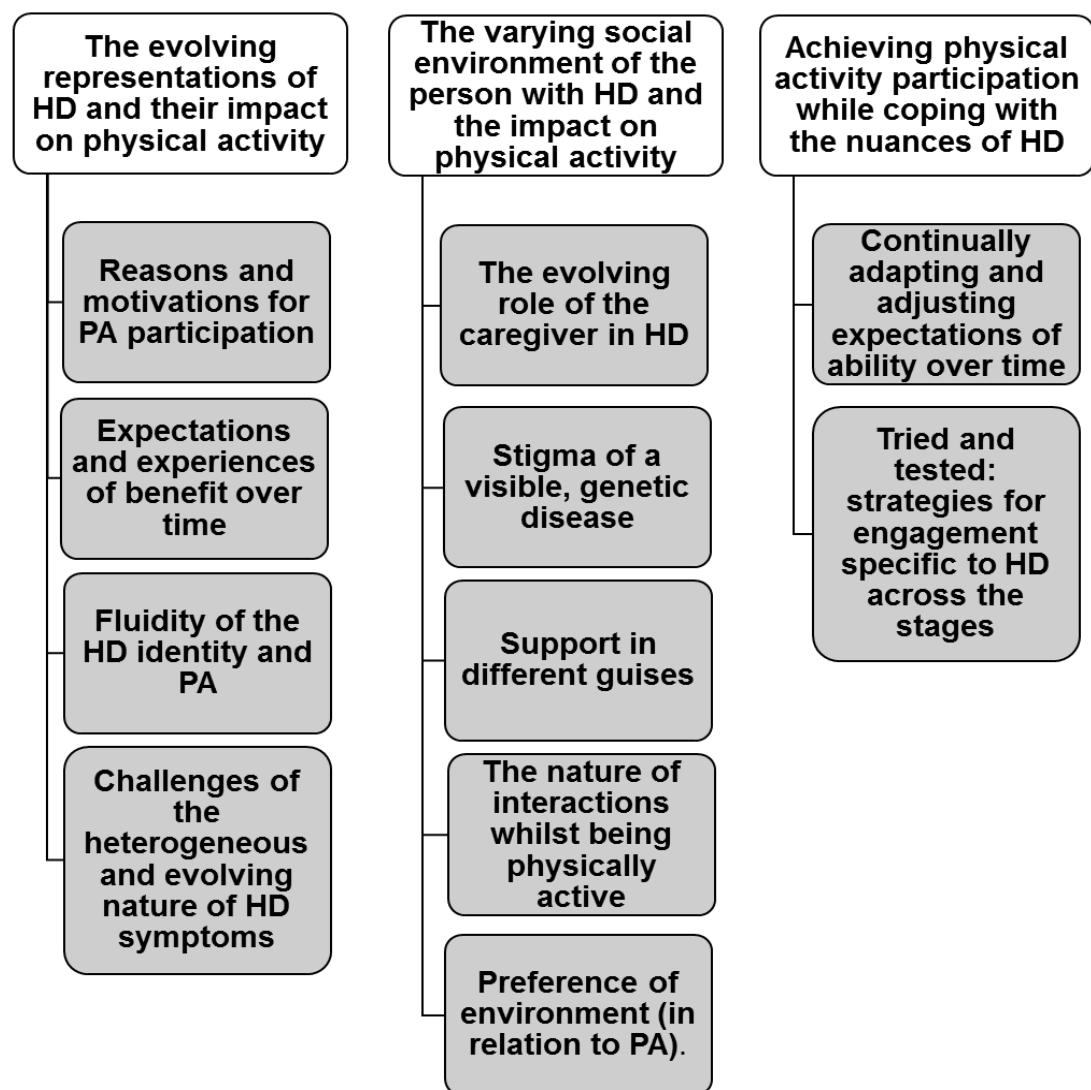
A rich and diverse picture of physical activity (PA) experiences in the context of Huntington's disease (HD) were elicited through in-depth analysis of the FG transcripts. Charting and mapping of data indexed to analytical coding framework which was developed using components of the Self-regulation model (SRM) and open coding of the data led to the development of **key themes** and subthemes (see figure 14). These themes describe and add to the existing knowledge about how the nuances of HD impact on PA. The three key themes derived from the analysis are namely 'the evolving representations of HD and physical activity', 'the varying social environment of the person with HD and the impact on physical activity' and 'achieving PA participation while coping with the nuances of HD'.

Theme 1, the 'evolving representations of HD and physical activity' encompasses the recurring concepts throughout the FGs of the changing nature of HD and how that impacts on PA. This includes the challenges and expectations and experiences of PA over time. The concept of illness representations within the SRM are relevant to this theme. Illness representations changed and evolved over time at the different stages of HD, there were changes in the challenges and expectations of PA engagement. Clare and Harman describe uncertainty of people with early stage dementia in the context of exploring illness representations of early stage dementia (Clare and Harman 2006). Linking with this it was apparent that the heterogeneous nature of HD challenged people in different ways over time. Key theme 2 is 'the varying social environment of the person with HD and the impact on physical activity'. The role of the social environment is seen in terms of preference, support and stigma for the people with HD in the context of PA participation; in particular, the crucial role of caregiver(s). The SRM describes direct social communication with others and the surrounding culture as a source of information for illness representations to develop (Levanthal et al. 1984). The caregiver influence in previous studies exploring experiences of HD has elicited the effect of their cognitive representations on the well-being of the individual with HD (Helder et al. 2002b; Kaptein et al. 2007). Linking in with the idea of social communication being a source of information for illness representations, (Levanthal et al. 1984) the caregiver role was also highlighted by the findings presented here.

Key theme 3; 'achieving physical activity participation while coping with the nuances of HD' comprises the concept of coping in the context of HD and PA. This relates to people with HD using PA as a way of coping, how coping or non-coping may affect PA behaviour and using strategies (caregivers and people with HD) to help maintain PA participation. Key to this theme is the idea of adapting with the disease progression.

This is suggestive that the participants engaged in an ongoing iterative cycle of processing information, to coping responses (adapting PA to suit their abilities), to appraisal, described as the integration of information by the SRM. Indeed, the Self-regulation model is a useful model in facilitating understanding of the impact of the nuances of HD on PA. The key and subthemes can be seen in figure 14 below. Figure 16 illustrates where the key themes identified from the data analysis sit in relation to the SRM constructs.

Figure 14: Diagrammatic Representation of key themes and subthemes



The SRM was used as the theoretical underpinning for the theoretically driven part of data analysis. A priori codes were developed from SRM constructs to contribute to the codes of the analytical coding framework (which also consisted of codes developed from open coding the data) and data was coded to them. As such, exploration of the

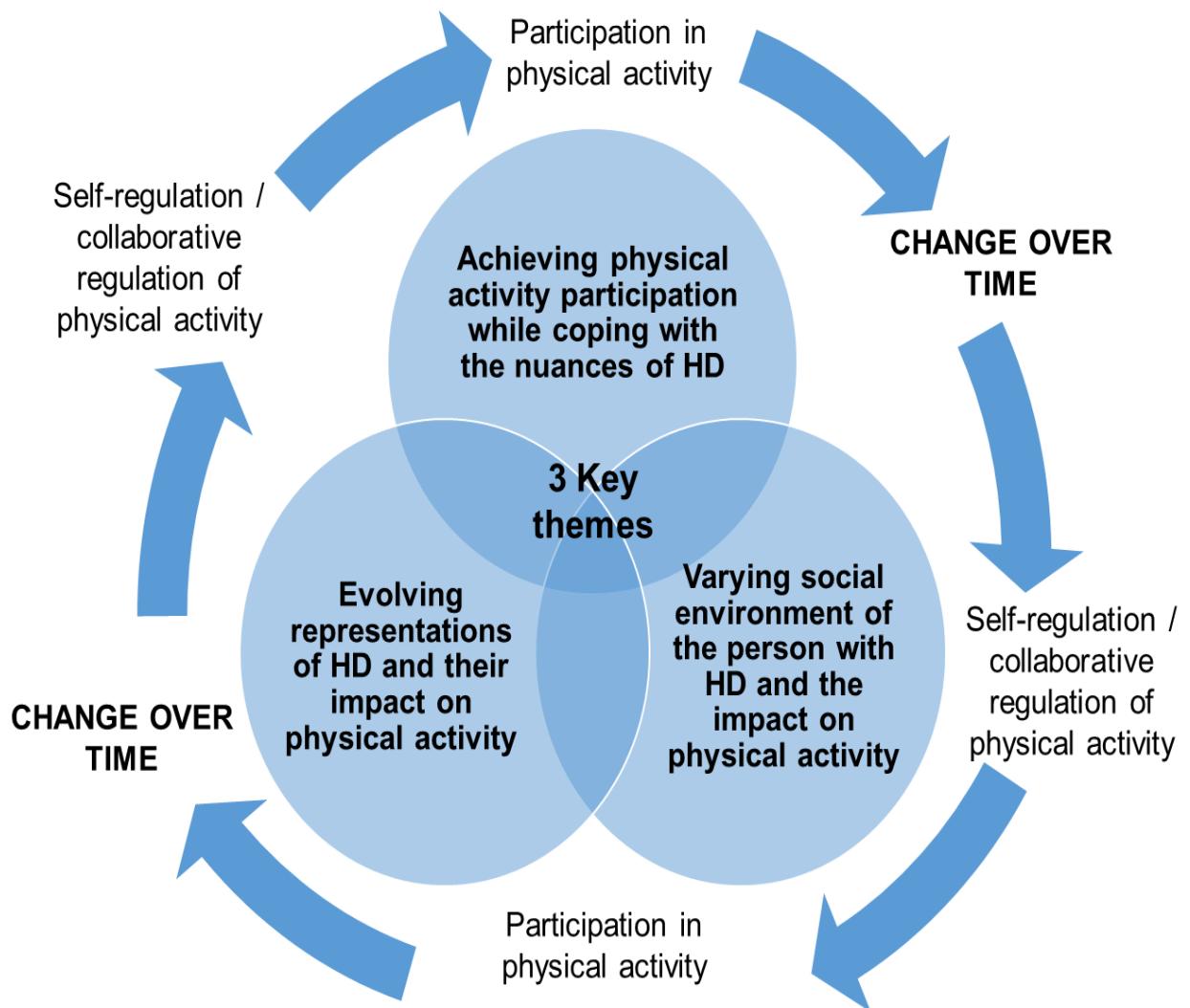
potential for the SRM to further understanding of experiences and perceptions of people with HD and their caregivers in relation to PA and the impact that the nuances of HD have on participation was facilitated. During analysis, a substantial amount of interesting and relevant data was coded to the a priori codes that were developed from SRM constructs, mainly ‘coping’, ‘consequences’, ‘control’, ‘identity’, ‘timeline’. This suggests that the constructs of the SRM were relevant to the constructed participant experiences and perceptions shared in the FGs. Discussion of how the key themes and sub-themes link to the SRM is given within presentation of each key theme and within the discussion chapter.

Another interesting finding that adds to the model is that with HD progression over time, caregivers play an active role in helping appraise and adapt PA participation of the individual with HD. Figure 15 illustrates an ongoing cycle where change over time is an ongoing issue in HD and in consideration of the overarching concept of PA participation in HD, self-regulation becomes more of a ‘collaborative regulation’ in monitoring success in an activity and adapting and changing activity plans. The key themes are surrounded by the ongoing cycle of participation in PA and regulation (self/collaborative) as these are the key concepts that influence and were found to be pertinent to the experience of PA in HD. Figure 15 also depicts the three key themes as overlapping because the concepts encompassed by each key theme are linked, they do stand-alone but they are not mutually exclusive. The terms ‘physical activity’ (PA) and ‘exercise’ were often used by participants interchangeably, since exercise is a type of PA, however there was some recognition of exercise being planned and structured by participants at the earlier stages of HD as in the example below.

Person with early stage HD, FG8:

*“I suppose what defines it from any other physical activity is whether it’s controlled...so, regime. Doing it at a certain time. So it becomes a routine...controlling. You know...so you can get in a certain amount of exercise whereas other things it’s just incidental.”*

Figure 15: Key themes in relation to the context of physical activity participation in HD and the interwoven thread of 'change over time' and self/collaborative regulation.



'Change over time' was identified as a major thread across the key themes in terms of what the changes in presentation of symptoms through the stages of HD mean for participating in PA. 'Time' was observed as a component of illness representation within the SRM and was used to develop the analytical coding framework. The findings suggest that 'time' (specifically for HD, 'change over time') has a key role in illness representations in relation to how those representations affect ongoing PA behaviour. Given the progressive degenerative nature of HD observed from previous literature and witnessed by the researcher in working with people with HD, it is perhaps natural to expect that there will be consequences for engaging in PA. The findings from the FGs concur with this perspective. The findings give insight into the nuances of living with symptom progression and the consequences for PA in terms of fears or apprehension and functional realities that people face over time.

Although there were negative influences of HD on PA which may seem to paint a bleak picture for people with HD, there were instances of positive experiences and perceptions of PA. Examples of overcoming barriers were described and ways in which people had adapted to positively influence their PA participation. This also in turn positively impacted on their perceptions of control; pertinent in a disease that takes control over body and mind and has no cure but people have to live with for a long time. Instances were discussed where participants evaluated how HD symptoms and their abilities affected their PA, and how they had adapted to be able to participate in PA. This suggests that the process of developing cognitive representations which guide actions and that lead to appraisal and evaluation and modifying activities (described by the SRM) is of relevance to PA in HD. With progression though, the regulation of behaviour may become more collaborative rather than entirely self-regulated as caregiver input becomes so vital to the daily life of the individual with HD.

#### **5.4 Key theme 1: The evolving representations of Huntington's disease and their impact on physical activity**

This key theme captures insight into the various perceptions of physical activity (PA) for people with HD across the spectrum from prodromal to late stage of the disease. The key themes and sub-themes were developed from mapping and synthesising the data indexed to the analytical coding framework as described in the methodology chapter. Links to the illness representation constructs of the SRM (cause, control, consequences, timeline, identity) are highlighted in the following sections that discuss the sub-themes of this key theme. The term 'evolving representations of HD' relates to the constructs within the SRM that are suggested contribute to the formation of cognitive representations; identity, coping, cause, consequences and control (Levanthal et al. 1984). The term 'evolving' refers to the changes over time evidenced from having participants at different stages in the FGs but also individual experiences of progression of HD and how that impacts PA. The sub themes emerged from the process of reviewing the data coded to the analytical coding framework, and making links across codes that were relevant to each other, and representative of a large number of participants. The four subthemes comprise: **reasons and motivations for PA participation; expectations and experiences of benefit over time; fluidity of the HD identity and PA; and challenges of the heterogeneous and evolving nature of HD symptoms.**

## **Reasons and motivations for PA participation**

Participants reported numerous reasons and motivations for PA participation. The FGs highlighted importance of caregiver involvement in supporting the PA of people with HD (even if further on in the disease). Given this, it was interesting to separate out the perspectives of each; regarding why people with HD keep active and why caregivers support them to be active or not. Different perspectives may have potential to be a cause of conflict between caregivers and people with HD but this did not emerge as a salient issue. The beliefs or attitude of the caregiver may affect the PA of the person with HD. This appeared to be the case for a number of the participants; in a positive way that encouraged participation, mirroring the findings of Khalil et al. (2012). On the contrary, in three cases, the strong exercise beliefs of a person with HD inspired their caregiver to be more active. For example:

Spouse (caregiver) FG7:

*[...] he's always encouraging me. And I do...well I don't like the idea of going out and doing it but actually I enjoy it. I feel good and happy in myself once I've done it. Erm, but I need a lot of prompting for that (laughs)...the thing is, mainly because I'm so busy.*

Reasons for being physically active included enjoyment, (which was mentioned most frequently), being outdoors, goal oriented towards maintaining activities associated with roles within the family, to improve functional abilities to be able to partake in other activities such as going on holiday, training to participate in a half marathon for charity. Other reasons included to feel normal, for social interaction, liking how they felt following sessions of PA, to 'fight back' against HD, that it was important to do because of the benefits they felt it had for aspects of the HD. The following quotes illustrate some of the reasons mentioned:

FG1:

*I do exercise on the bike cos I like to be outside enjoying the exercise. Going outside, getting some fresh air.*

FG5:

*I like it. Love it...it's fun...I love going out [dancing]*

FG8:

*The black cloud in the morning isn't motivating but you must get rid of it somehow...and on the treadmill you can [...]. It's just a feeling of total negativity and that's my main motivator into physical activity. [...] It's one of those things that I think is very important. And I think exercise is, in a number of areas. Mental and physical. And I think as I have HD and from reading about the subject of HD it's important to do both...physical. My exercise regime...I find if I*

*don't do my physical exercise I get really bad. If I can do exercise every morning and release some endorphins it helps considerably with the Huntington's."*

Comparisons with the existing HD literature about PA experiences illustrate that facilitators to participate in exercise were similar. The broader exploration facilitated by the FG discussions elicited more information than previous studies of specific exercise interventions in HD (Zinzi et al. 2007; Busse et al. 2013; Quinn et al. 2014). Outcome expectations were important facilitators for a home based exercise programme for people with mid-stage HD (Khalil et al. 2012). Outcome expectations included that taking part in the exercise programme would help participants manage their condition and self-efficacy in terms of confidence in their ability to use the exercise DVD and do the exercises properly (Khalil et al. 2012). The idea of 'goal relevance' is described by Levanthal et al. (1998) in relation to the SRM, whereby the selection of response is perceived to have direct access to the targeted health threat. An example of goal relevance and outcome expectations elicited during the FGs of this research is of a participant with HD, a female at mid to late stage in FG3. Supported by her caregiver, this participant set functional goals to improve her functional abilities that had declined because of HD and inactivity. The participant had the expectation of being able to go on holiday for the first time in a while by improving her functional abilities through PA. In this case, outcome expectations could be linked to goal setting. By setting the goal, the participant expected to work towards that goal with the aim of achieving it, and therefore had positive outcome expectations. This is reflected in a quote from the participant (and her caregiver adds some context to the quote):

*"Because I'm aiming to go on holiday aren't I..."*

Caregiver:

*"[name]'s motivation is being if her mobility increased, which it has through the exercises she's been doing, she's achieved that, then her husband would take her away with the family and they'll go to [location] now for four weeks now and over to to their place in [location]"*

Positive outcome expectations and goal setting appeared to influence PA behaviours for FG participants. In light of this, concepts such as outcome expectations and goal setting described by Bandura's self-efficacy theory (1993) are relevant to the findings of the FGs. This links to the overarching key theme of 'evolving representations of HD and their impact on physical activity' because over time, disabilities will become more prominent. This may impact negatively on self-efficacy and positive outcome expectations and the individual's cognitive representations of the disease. In addition to

perceived confidence to carry out activities being important for adherence in Multiple Sclerosis (MS) and Parkinson's disease (PD) (Morris et al. 2008; Ellis et al. 2011; Ellis et al. 2013), Smith et al. (2009) found that confidence in ability to exercise safely within their limitations caused by the disease was an important factor for people with MS. A similar example from the FGs is given in the quote below:

Spouse of woman with early stage HD, FG4:

*"(Physical activity regime)...helping [name] to cope with the disability and to feel confident about movement and exercise within the disability and the environment, so that there's a confidence that she could have to remain independent and that's the thing isn't it?"*

The spouse explains the need to help improve the individual's confidence. This is in terms of doing activities in the correct way or feeling able to actually do the activity to remain independent despite the disability caused by HD. This was reported by caregivers of people from early to mid-late stage HD in the FGs.

From the caregiver perspectives, enjoyment of PA was highlighted as a reason for the person with HD being physically active. A caregiver summarises in FG1:

*"It sounds like exercise has got to be something that people enjoy"*

The generally positive attitude towards PA of FG8 participants is reflected unmistakably in the exchange between 3 participants with HD below where they are discussing their reasons for being physically active:

P5:

*"And the lifestyle changes...like...life style changes for me also...I don't drink as much as I used to...Not that I was ever drinking a lot!...and that feeling you get from exercise afterwards...released endorphins. It's really powerful. It's can be addictive."*

P1:

*"It's definitely addictive. It is isn't it! If you miss...you know. If you haven't been exercising for a couple of days because maybe you haven't been that well. You do start to feel like you want to get back into that stretching and limber feeling."*

P8:

*"I agree with that totally. You should really try and exercise everyday if you can...I think once it's released endorphins I just feel so much better... I guess I used to do lots of running and it does make you feel a lot better...less stressful."*

Perhaps experience of seeing relatives deteriorating motivates this gene positive person in FG8:

*[...] you will lose it if you don't use it so that's important. He's got a Nintendo Wii now so that we can get him to play tennis and try and build it back up again but if you sit there and do nothing you will suffer...he's visibly in pain."*

This certainly resonates with the findings of Quinn et al. (2010) who found that a facilitator to being active for people with PD and HD was the perception of exercise being beneficial for managing the symptoms of a movement disorder. In addition to the findings by Quinn et al. (2010), retaining the 'healthy self' was a strong motivator to PA for people with PD by Eriksson et al. (2013) who interviewed participants of an exercise programme for Parkinson's disease (PD). It may be helpful in the context of HD to perhaps consider retaining the healthy self for as long as possible because people live with the symptoms over such a prolonged period of time. 'Retaining the healthy self' is suggestive that it is possible to stop progression which would not be a true reflection for HD. Rather PA participation may help maintain physical function for longer by slowing down symptom progression or moderating symptoms. For some FG participants, the concept of managing HD through PA appears to be interwoven with the idea of 'taking control' and perceived benefits of PA. To capture the feeling of taking control as a motivator for PA is certainly portrayed strongly by particular participants. For example:

Person with early stage HD, FG8:

*"The black cloud in the morning isn't motivating but you must get rid of it somehow...and on the treadmill you can [...] help you fight back and do something."*

Talking about fighting back suggests that this participant feels strongly about his role in dealing with HD. People with PD have spoken similarly about the disease that they describe takes over their bodies when relaying their experiences and perspectives of PA (Ravenek & Schneider 2009). Parallels highlighted in perceptions of PA as a means of control or trying to take control as a motivator may be because like HD, PD is a long term disease i.e. people know that it is progressive and want to do something to delay progression for as long as possible. Certainly in the previous literature relating to representations of HD, people with HD and their caregivers characterise it as a disease of long duration with lack of cure or possibility of symptoms improving (Helder et al. 2002a). They have also expressed feeling little control over the disease in a personal sense and in terms of treatment (Arran et al. 2014).

The purposeful attitude of engaging in PA to take control over their lives appeared to be relevant to those at the earlier stages of HD. Although the lines between maintaining control, maintaining independence and how these goals motivate individuals were often blurred. People were clearly trying to hold onto a sense of control over their lives and a sense of personal agency in what they do. For some this meant battling along trying to continue doing what had always been normal for them. For others it was embracing and taking decisive action in terms of adapting their PA to make it more achievable and realistic as per their decline in abilities. Consistent with the findings by Arran et al. (2014) the effort to maintain control was sometimes described in a way suggestive of a constant struggle. It may be that although people strive to maintain control they may not necessarily actually feel in control. For some it was about coming to terms with the progression of the disease being out of their control. The HD was something that was happening to them rather than something that was a part of them and they did not see or accept it as an integrated part of them, part of their 'self'. The terms 'denial' and lack of 'acceptance' were used a number of times by caregivers which ties in with this concept. A striking example was shared by a spouse, whose husband (FG2) who still attended his local golf course and was taken around by friends who were members, despite him having profound physical difficulties.

Caregiver (spouse), FG2:

*[...] a lot of Huntington's Disease is denial of perhaps of what's going on and by carrying, by doing the things that he done before he was denying that anything was wrong... my husband played golf of some description right up until the time he was in a wheelchair and we had a tiny little 9 hole course just down the road from us and a couple of men down there were just really really kind to him and would take him round and I don't know quite what he did (laughter)... he certainly couldn't fold his trolley down to get it into a car and he certainly couldn't do a shoe lace up if it came undone ..."*

Another caregiver in a different FG (6) explains that his wife in the later stages still wanted to be mobile although it was no longer safe and she needed to be in the wheelchair. The quote from the caregiver below suggests awareness of the tension for the person with HD between the subjective experiences of wanting to maintain control versus the objective reality of lack of control over the progression of the disease.

Caregiver (spouse), FG6:

*"I don't think she appreciates the fact ... she's got no appreciation. She has trouble accepting, accepting the fact that she can't do."*

Going out for regular daily walks was important for one woman with early stage HD who her husband had described as having some cognitive decline. They talked about how doing this was enabling them to maintain 'normality'. It appeared that they were still able to continue at this point and using PA as a means of control to maintain normality. There were poignant examples where people with HD talked about PA as a means to try and maintain independence. However, caregivers emphasised aspects of maintaining independence, daily physical function and trying to continue as normal for as long as possible, more as reasons for why people with HD were active.

A caregiver in FG4 explains:

*"I suppose the things that we've talked about there in terms of trying to maintain some sense of normality from doing those things is a big feature for [name] to go for that walk to be able to do it to be able to have done it is very important to you...it's involving some level of physical activity which can constitute as exercise...the longer we can kind of keep it in place, better off we're going to be."*

A caregiver gives insight into his spouse's motivation to do exercises. He explains that his spouse wanted to please the physiotherapist who had given her the exercises to do. He also hints at another aspect of HD which can be obsessive behaviour or thoughts (Walker 2007) and may influence PA behaviour. A reflective comment of another participant with early stage HD (below) is also suggestive of this.

Caregiver, FG1:

*"...while she was doing it there was a vast improvement and she'd got in that mind-set that (the physiotherapist) had left those instructions to do those exercises every day and she religiously carried them out so I think the HD once they get something in their mind n especially if it's that, then they'll do it"*

Person with early stage HD, FG1:

*"I suddenly realised that it was about 2 hours per day on the bike it's a bit more than an ordinary session of exercise."*

Another reason for PA participation was that it was a long term habit, part of a lifestyle that participants had followed for years before diagnosis of HD. For example, a caregiver in FG7 shared their point of view regarding their spouse who had been very physically active for a long time:

*[name] has always exercised. He used to do triathlons when he was younger and things like that so it's always been a big part of his life. And he likes the feeling of having...do you want to carry on? (Turns to spouse with HD)..of having worked out and the muscles feeling achy the next day. He enjoys that and the endorphins flowing and he enjoys that as a mood..."*

In contrast to the above participant, a caregiver from another FG volunteers their thoughts about the lifestyle of a young woman living at the assisted living facility at early to mid-stage who was affected more behaviourally than physically.

Caregiver, FG5:

*[...] she had no intention to, it's not like something she would have found natural thing for her to do. She wasn't one of them who'd go out and join the gym..."*

Taking into consideration the progressive nature of HD, if PA was an ingrained 'habit' i.e. part of their day to day lifestyle at an early stage, people may continue and adapt as needed as the HD progresses. It may be harder to initiate PA for somebody at mid stage, where there are more barriers because of symptom progression and the representation they hold of their HD by that point. For example, altered gait and balance and fear of falling in HD and PD has been attributed to lower functional mobility and reduced PA may be indicative of increased falls (Busse et al. 2009; Varanese et al. 2011). In addition, cognitive difficulties mean that recall of instructions may be difficult. In contrast, recognition is easier than recall for people with HD, (Pollard 2008). Recognition of how to do physical activities or specific exercises as demonstrated by instructors may trigger a person's memory of what to do if it was an activity that they had previously done and had memories of doing it.

In observing the literature regarding PA in PD and MS, most of the studies explore maintenance of PA with people who are active and current exercise beliefs or behaviours (Eriksson et al. 2013; Ellis et al. 2011; Kayes et al. 2011). The role of exercise beliefs or behaviour prior to onset of the conditions and how or whether that plays a role in engagement in PA following diagnosis does not appear to have been explored in previous literature regarding HD and PA. It is highlighted as a potential influencing factor for people with HD in the FGs. It may be more pertinent to explore

current beliefs but understanding previous PA behaviour and experiences may help to explore and address negative perceptions that could form barriers to PA.

Mainly similarities emerged between perspectives of the people with HD and caregivers in terms of considering the motivations or reasons for people with HD to be physically active. Enjoyment was portrayed as a motivator from early to late stage, and was highlighted by the caregivers. Maintaining independence was a salient issue and reason that both people with HD and caregivers gave for being physically active. Social interaction was also an important reason for people at different stages. An example of a shared perspective between a person with HD and her spouse (caregiver) is illustrated here from FG4.

Person with HD, FG4:

*„,it's a good feeling so yeh it's like um a good feeling at the moment I'll try and keep that up for as long as I can [...] obviously you get to know the ladies that go to the class as well so it's a social thing as well...and if you can do exercise it does put you on the same level as everyone else in the group.”*

Caregiver (spouse), FG4:

*“I think for [name] it's a big feature of why she exercises, walks the dog, goes to pilates because it's one of the few areas left in her life where she can actually interact with people outside”*

The importance of social participation for particular people in the FGs reflects findings by Eriksson et al. (2013) and Skår et al. (2014) in MS. Eriksson et al. (2013) and Skår et al. (2014) also found that the social participation of exercising with similar others was a facilitator to engaging in exercise. In the context of social participation, caregivers and some participants with HD briefly discussed their thoughts on exercising with others with HD. What came across more strongly for particular participants in the early stage was the ‘want’ to join in and socialise with ‘healthy’ people. There could be a number of reasons for this. People spoke about feeling normal, and exercising with ‘healthy’ people might be a part of that. Although what should also be considered is that there are not any exercise groups specifically for people with HD and people with HD tend to be fairly isolated. Therefore, it would be natural for them to only consider the context of exercising with ‘healthy’ individuals.

In considering motivations and reasons for PA in HD people seem to be motivated to continue being physically active for the positive benefits perceived to be gained because of the PA. Specifically this is relevant to the feedback loop of the self-

regulation model (SRM) where experiences can either reinforce or facilitate a change in behaviour following appraisal of the behaviour/actions and the consequences; positive or negative (Levanthal et al. 1984). This also certainly reflects observations of the researcher when previously supervising people with HD in their community gym or at home; that positive gains spurred people onto continuing with their exercise programmes.

Personal preference in terms of enjoyment and dislike was emphasised by a woman in FG3 with mid to late stage HD. Despite being an avid skier before her HD symptoms manifested, the gym had always ‘terrified’ her. It was not since developing HD that she felt this way, it had always been an intimidating environment to her. Regardless of stage of HD going to the gym is not something she would contemplate doing. Individual experiences of the HD and previous exercise or PA history may well have shaped perceptions of what they are able to do in terms of PA, but enjoyment or dislike were also influencing factors. This finding is in concordance with that of a qualitative study of PA in MS where people had not engaged in certain PA before developing MS because they did not want to and nor had they become interested since developing MS (Kayes et al. 2011). It was not that the MS had stopped them, they had just never been interested (Kayes et al. 2011). These findings were elicited through semi-structured interviews with 10 people with MS that sought to explore barriers and facilitators to engagement in PA (Kayes et al. 2011). Although not something unique to HD, consideration of enjoyment as well as capabilities as a key factor could be important for adherence to personal PA plans.

### **Expectations and experiences of benefit over time**

FG participants discussed numerous perceived benefits of participating in regular PA. Participants discussed what they believed were the benefits that *could* be gained and the benefits that they had actually experienced. Such discussions reflected representations operating at an abstract as well as an experiential level. Participants with HD discussed benefits of PA participation in terms of a better lifestyle and health, quality of life, maintaining independence, functional abilities, and the opportunity for social interaction; important in an ‘isolating disease’ as described by participants. For some, engaging in PA made *them* feel as though they were doing something positive for their health and gave a sense of taking control. This is something that may be particularly important in a progressive, long term disease for which there is no cure.

The comment from a woman with mid to late stage HD in FG2 suggests that going to hydrotherapy sessions used to make her happy, suggesting that participation in the

sessions had a positive impact for this individual. Although this participant struggled with her speech, she did manage to articulate this point succinctly;

*“I used to smile when I went in.”*

The caregiver went on to explain that unfortunately the classes were subsequently stopped by the service. A gene positive person in FG8 was training for a half marathon at the time of attending the FG. He talked about the positive effects that he perceived, even though it was not always easy to find the motivation.

*“...obviously your mind prevents you from doing a lot of things, particularly in terms of exercise [...] the lifestyle changes...like...life style changes for me also...I don’t drink as much as I used to...Not that I was ever drinking a lot!...and that feeling you get from exercise afterwards...released endorphins. It’s really powerful.”*

In relation to the perceived benefits of engaging in PA, a participant of FG8 gave a very clear example of what PA means to them, specifically in the context of the HD. He was an older man at the earlier stages, living with his wife who had always been active (a professional ballet dancer) and was noticeably an advocate of PA in HD.

FG8:

*“My exercise regime, I find if I don’t do my physical exercise I get really bad. If I can do exercise every morning and release some endorphins it helps considerably with the Huntington’s... The black cloud in the morning isn’t motivating but you must get rid of it somehow...and on the treadmill you can.”*

At this point the researcher probed further, asking ‘what is the black cloud?’ to which the participant responded and an interesting exchange then followed between participants:

Person with HD:      *“Anyone with Huntington’s would probably understand it exactly. It’s just a feeling of total negativity and that’s my main motivator into physical activity...”*

Caregiver (spouse): *“Is it after the exercise that you feel the benefit?”*

Person with HD:      *“Oh, absolutely.”*

Caregiver (spouse): *“Or shortly after you’ve got on the treadmill or after you’ve finished?”*

Person with HD:      *“It’s basically comes during. About half way through I mix it up. I do twenty minutes usually...about ten minutes I begin to feel that the world is ok again. That nasty cloud is gone”*

Caregiver (spouse): “*Amazing.*”

Gene positive person: “*I think that shows how important exercise is...*”

Person with HD: “*It is...to anybody. Obviously with Huntington's to help you fight back and do something.*”

Other participants in the group seemed to take note of what this person (he) said. He perhaps highlighted a different motivation for PA; to feel a positive sense of control or of doing something to challenge the feeling of helplessness that HD can cause. This ties in with the concept of motivations and reasons to exercise. Again, it echoes findings of studies in MS and PD where physically active participants disclosed perceptions of ‘fighting back’ against the disease (Ravenek & Schneider 2009; Plow et al. 2009) Indeed Plow et al. (2009) found that active participants with PD felt they had control over the disease.

Regarding the physical benefits of PA for retaining function, the element of time and change over time was apparent in the thoughts of participants even at the earlier stages. This anticipation seemed to be linked to seeing other family members with HD. For example, the gene positive person in FG8 who had seen the progression of HD in his father explained:

*“My dad [...] one of the things he's noticed is that because of lack of physical activity he's lost muscle definition and stuff like that...so if you don't use it you will lose it. Basically he can't throw a ball at me now without being in some form of pain so basically if you just sit there and do nothing you will lose your muscle. So you know, just trying to get him to interact with the ball and keep him moving. He's losing muscle definition simply through sitting around and inactivity...”*

Caregivers shared observations of physical improvements in strength and stability of the person with HD through engagement in PA. They also noticed improved mood, functional abilities, improved confidence, quality of life, and retaining a sense of normality through PA participation.

Caregiver (spouse), FG7:

*“...he likes the feeling of having...do you want to carry on?...of having worked out and the muscles feeling achy the next day. He enjoys that and the endorphins flowing and he enjoys that as a mood lifter”*

Caregiver (spouse), FG7:

*“It seems to keep [name] a lot less wobbly whereas before when she wasn't doing as much exercise her movements were greater but now her core is a lot*

*stronger she's a lot more in control. She used to bump into things a lot more, bumping into frames and things but now she's spot on. Whether she's actually able to correct herself a bit quicker I don't know but it's making her a lot more stable [...] she'd bash her head on the door frame. Things like that, but you know. She hasn't done that in a long time [...] now things are getting a lot more stronger she's definitely in more control. I think [name] can correct herself a lot quicker because the strength is there..."*

As well as being a motivator to PA, social interactions and support were indicated as benefits of taking part in PA. An example of an older man in FG2 with early stage HD shares his perception of the 'social aspect' as being facilitative of PA for him:

*"...you've got a social obligation you know I can't let my friend down so I've got to make the effort, so it's a social obligation that you felt and also enjoyed doing as well that that social obligation to meet up with other people to do..."*

Exploration through the FGs across the spectrum of HD from prodromal to late-stage has highlighted the importance of social interactions and social support across the stages, and the potential benefit of support for self-efficacy in PA. The findings of Zinzi et al. (2007) suggested that social support provided as part of an exercise intervention for people with early to mid-stage HD was perceived to improve well-being. In relation to the wider literature of other chronic and neurological conditions, social support has been found to be an important influence on PA participation (Motl et al. 2009; Ravenek & Schneider 2009; Learmonth et al. 2013).

The spouse of a young man in FG8 observed that seeing positive differences in their appearance was a motivator for their training to complete a half marathon.

Spouse of gene positive person:

*"And also, [name] obviously as a result of going [running], his body shape is changing, so as that's started to happen that's quite motivational isn't it that sort of feeling of, it's not so much the scales or the scales aren't so important, definitely"*

### **Fluidity of the HD identity and PA**

Various discussions were elicited that formed a thread around the concept of identity. Role identities within the family related to PA, how the changing identity of HD changes exercise identities were discussed. HD participants also shared their experiences of how HD is perceived to be something that masks the identity of an individual so that

outsiders only see the HD. These concepts incorporate aspects of how people label HD, not just what the symptoms are, but what the symptoms mean for their individual identities linked to PA. The relevance of the SRM is highlighted here because how people label HD does appear to affect people's perceptions of and motivations for PA. For example, regarding role identities within the family, HD was perceived to be something that forces a change in roles and dynamics within family units and threatens 'normal' family life (expanded upon later in this subtheme). In the context of PA, this threat was met by some participants using PA to try maintaining function, in order to maintain activities associated with their family roles. The identity and labelling of HD emerged as something that changes over time as symptoms progress, as seen in the differences between people at different stages in the FGs, hence the subtheme title of 'fluidity of the HD identity'.

Across three FGs, the roles of women within the family unit were discussed (about women who had HD). Role identities of 'wife' and 'mother' emerged in the context of retaining activities associated with these roles (by the participants) by trying to maintain function through PA. For one woman with HD her caregiver said that she constantly reminded the woman with HD that she was a wife and a mother. The caregiver encouraged her to maintain a routine that included shopping, collecting prescriptions and baking with assistance. A succinct quote explaining this was given by the caregiver:

*"I kept saying to her; you're a wife, you're a mother, you're a woman."*

The daily activities described above were all part of her PA plan following input from a physiotherapist where functional goals were agreed with the woman with HD and the caregiver.

*"You need to become part of the family again because her husband and everybody was doing the shopping and everything for you. And now [name] does the weekly shop herself, umm, she goes out, she collects her prescriptions, everything that she needs that now has made her become a wife and a mother...again, which she wasn't having before, when we first started."*

A female participant with HD who attended FG4 with her husband also talked about maintaining her role within the family unit. The couple were in their thirties and had young children. The diagnosis was made four years prior and the woman had previously overcome cancer only to then be diagnosed with HD. They were coming to terms with living with HD. The woman was at early stage and they disclosed that she was experiencing cognitive symptoms such as memory loss and manifestation of motor

symptoms and balance impairment. The couple and their children were isolated from the side of the family where HD had emerged because the other family members did not want to acknowledge the HD. The husband explains this issue:

*“...knowledge about people in the family suffering from it is the biggest problem. That’s partly part of why the family has come apart, because we are open and honest about it and they are not. They don’t let their kids see us, they don’t let our kids interact with one another so on and so forth. That’s that, yeah, that’s that side. My sisters. You’re not allowed to mention Huntington’s.”*

The woman with HD and her spouse seemed to make a link from their perspective between maintaining her walking, being able to get to the shops independently and being able to provide for her children. The word ‘try’ possibly suggests this is not always doable. In addition, ‘trying to maintain a sense of normality’ suggests their awareness of the potential for HD to affect what they consider to be normal family life.

*“...trying to maintain some sense of normality from doing those things is a big feature for [name] to go for that walk to be able to do it to be able to have done it is very important to you.”*

For women with PD, the desire to normalise the disease rather than denying that it existed was linked to the aspiration of being ‘good mothers’ (Fleming et al. 2004). In both PD and HD, normalising a disease where so much of daily living is progressively affected would likely be decreasingly feasible.

From another FG, the partner of a female with HD suggested that being active was a big part of the participant’s role as a mother and wife. Although it is very difficult, she still wanted to try to walk and stand up to do things. The caregiver may consider her differently because of the changes caused by HD perhaps because she can no longer complete functional activities associated with roles that they traditionally relate to being a wife and a mother. The example below is taken from a caregiver where he talks about his wife and how the changes due to HD have affected her ability to continue the activities she used to carry out.

Caregiver, FG6:

*“I think if you’ve been a wife and a mother you are on the go all the time and it must come as a big shock”*

Participants talked about changing roles as well as ‘maintaining roles’. For example, spouses took on more caring roles as the HD progressed. HD is labelled as something that takes away role identities over time from early to later stages. For example:

Caregiver, FG6:

*“she’s been active for so many years and then having to rely on myself...you know, it gets to her, why sort of thing, sorry I can’t help you... during my illness I say she’s been there and helped me, say it’s my turn now, innit hey... she did fight to stay on her feet.”*

Throughout the FGs there was a mixture of people with strong exercise identities, and lesser so. For some, exercise was an important part of them and their lifestyle. For those participants whom PA was an ingrained, regular part of their daily life, the want to continue was still present for most and they wanted to retain a strong exercise identity as such. Some continued but adapted what they did (with support) because of the limitations caused by HD. While for others the symptom progression had caused a complete change in what they were capable of. In their view, the consequences of HD presented limitations to participation, portrayed in the quote here:

*“I used to do all those things a real keen sportsman; football, cricket but this illness means that I’m not steady on my feet.”*

An example of adapting is illustrated by a person with HD:

*“...you know I used to be very active in the navy and now that has dropped because of Huntington’s..., I run around as much as possible...just trying to get out of the wheelchair as long as I can”*

In FG4 a young man in his twenties (quoted below) was residing in an assisted living facility. For him, the functional consequences of HD had meant adapting his PA.

*“Exercised all my life, football I played, rugby I played, cricket I played, half marathons...now doing sit ups at my mum and dads the weekend...football we play every week [football in the activities group at the assisted living facility] ...Yeah, every week, badminton we play [he went to the group exercises at the assisted living facility on Friday].”*

Here a caregiver talks about the woman that she cares for at the later stages of HD who can no longer continue what she used to do:

*“...the drive is still there and you know, the passion’s still there that she enjoys running but because of her mobility [she no longer runs]”*

Another person with HD in FG6 expresses their frustration at their physical limitations caused by HD because they used to be more active:

*"The frustration that you can't do it but you want to do it.*

The above quote illustrates the consequences of HD for this particular participant. The issue echoes findings by Fleming et al. (2004) where women with PD described their grief at having to change their PA because of limited abilities but still experiencing the impulse to want to do it. An example is given here of somebody who could not continue to play golf but still attended the gathering at the golf course. Perhaps the social aspect was important to them and to retain those social links, although not physically capable of participating:

*"... he certainly couldn't fold his trolley down to get it into a car and he certainly couldn't do a shoe lace up if it came undone but the people he played with were just so supportive and the family who run the golf course were really understanding and so he had a really good run for his money...you know he done things a lot longer than most people would probably be able to"*

Amongst people at earlier stages of HD, there was a common perception that they look physically different or move differently, but would like to have social acceptance.

Person with early stage HD, FG8:

*"...and I kind of noticed, you know, I'm not the same as everybody else"*

Woman with early stage HD, FG4:

*"...it's good to um be social and be with other people, interacting as well as you can you know? To a point where they stop noticing things."*

Participants across the groups talked about HD as 'masking' the identity of the individual because people could not see past their visible motor symptoms. Vexation at this was expressed by both people with HD and caregivers because of the consequences for how others perceived them. One caregiver in FG7 seemed almost pleading in sharing their experiences of this:

*"Don't just see them as an HD sufferer"*

An example is given from a couple who attended FG4:

Spouse / caregiver of woman with early stage HD:

*"I think most people when we walk along think she's drunk"*

*Woman with HD:*

*"Yeh cos I'd be drunk at 11 o clock in the morning!"*

*Spouse / caregiver of woman with early stage HD:*

*"...the characteristics are uncomfortable for people to deal with so they tend to say 'oh that's [name] she's a bit odd' you know"*

It is clear from the above quotes that the social identity of people with HD was something of a concern. Caregivers and people with HD discussed the need for awareness in others that they are *people with HD* and that HD does not define them as a person. Another frustration highlighted by some caregivers and people with HD was that health issues unrelated to HD were overlooked by healthcare professionals because it was assumed that all problems were a result of the HD.

*"They need to be seen as an individual who has HD and HD is XYZ, but this is the person's needs"*

*"It's...others also assume very often when they hear about it, what it is. I call the Huntington's a ghost. Because every symptom you develop...everyone immediately sees the Huntington's"*

Related to this, the possibility of other co-morbidities was highlighted by this caregiver in another FG:

*"goes to a nursing home twice a week for day care and on a Monday she gets weighed....and I keep an eye on her weight ahh... she's been pretty stable for last couple of 12 months but the only thing now, we've got that I didn't know about was (low) blood pressure, I've got to keep an eye on that."*

Although in the analysis, the code co-morbidities was deleted because of the lack of data it represented, the issue highlighted can have implications for PA. It is important to recognise that people with HD may have other conditions too and that HD should not mask diagnosing these, as argued by the participants. Recurrent across the FGs, family members discussed 'characteristics' of people with HD, and how these characteristics impacted on their PA. These characteristics were a result of the physical, cognitive or behavioural symptoms of HD

*"sometimes you have to accept it and not push, cos that's the nature of her illness...in time she'll make that choice for herself, and when she does no one will get on that bike!"*

Although caregivers/family members wanted people to see beyond the HD, in a way, they labelled the people with HD too, as they would generalise from their experiences, how people with HD ‘are’. For example, saying they can be ‘stubborn’ but this might be due to behavioural consequences of HD where people with HD like routine and don’t like to change their routine. Or labelling people as ‘lazy’ where as it could be apathy, part of the pathology seen in HD, which causes lack of motivation. In a way they are in some cases defined by their symptoms (through perhaps subconscious labelling) on the caregiver’s part.

A caregiver in FG5 explains their perspective:

*“...generally people with Huntington’s they’ll get something in their head and once it’s in their head it will stay there. So spontaneity doesn’t work most of the time...”*

### **Challenges of the heterogeneous and evolving nature of HD symptoms and barriers to PA**

The consequences of HD are significant to this subtheme and tie in with the SRM. Illness representations of HD and perceptions of the consequences of the disease are seen to contribute to formation of individuals’ cognitive representations of PA. The analytical coding framework codes that were related to the category ‘consequences’ were combined to form this subtheme. These codes were ‘impact of symptoms’ (impact of HD on physical, social, psychological function); ‘continuity’ (perspective of how life will continue (also in terms of PA) in the context of the consequences of HD); and ‘quality of life’ (perspectives with regard to quality of life, how is it altered? Does PA mean anything in terms of improving or maintaining quality of life?)

In comparison to the wider literature, perceptions of PA having a negative effect on symptoms (e.g. fatigue) has been found to be a barrier to PA for people with MS (Kayes et al. 2011). This perspective was not elicited from the FGs. Although people discussed arranging their PA around times when people knew they would be tired, PA was not considered in a negative light in terms of having a worsening effect on symptoms. This could be due to the difference in symptoms between MS and HD, or it could be that people with negative experiences did not want to attend the FGs to discuss exercise. However, for the groups of people that did attend which included prodromal to mid- late stage HD across eight FGs, negative perceptions of possible

effects of what PA might do to the disease symptoms was not a salient issue as with MS.

Perceived barriers related to PA participation specific to HD were mainly linked to illness representations of the physical, cognitive and behavioural symptoms. In the study by Kayes et al. (2011) a ‘cognitive tension’ between exercising and the perception of exacerbating symptoms was elicited. This appeared to be vital to participants’ decisions to engage (or not) in PA. In some cases, caregivers feared that PA put people with HD at increased risk of falling. For the most part though, the focus of discussion was the negative impact of HD on reducing participation rather than exercise having a negative effect on HD.

It was apparent from discussion within FGs and comparing data across FGs that manifestation of symptoms is different for different people and therefore the nuances of the barriers that they experienced were also varied. A clear example of this is given by two caregivers (who were spouses) in FG2.

Caregiver of person in the FG:

*[...] really I suppose with [name] it's limited physically but she's mentally very very good, she, my, she got a better mind than I have, my memory is shocking"*

Caregiver of person in nursing home, FG2:

*"For my husband...manifested in a slightly different way from [name of FG participant], he was actually far more mentally affected and behavioural problems more than physical but you know he did have some physical symptoms."*

Caregiver of person with mid to late stage HD, FG2:

*[name]'s brother and sister's got it, her brother has passed away now and the all three have got something different, there's not one thing in common, well there are one or two things that are common to them all like the involuntary movement and all that, but speech is different for some, the mind is effected more with others, so each individual have got so many different symptoms that is not one common thing, so a group of 8 or 10 Huntington's sufferers is like a 10 mile radius that get together once a fortnight or once a month, could the do the same thing anyway together because of the diversity in...what's the word, you know what I'm getting at."*

To illustrate the HD symptoms reported as causing barriers to participation, the physical, cognitive and behavioural symptoms have been highlighted as separate entities for ease of reading. Reflected in the example below, the researcher acknowledges that in reality the symptoms of HD do not occur in isolation, but not everybody necessarily experiences all of the symptoms equally either. In turn, how this

impacts on PA participation is also different for individuals across the stages and within the stages of HD.

Caregiver (spouse), FG4:

*"So the exercise thing I think has kind of become more constraining, there's two things that do it one is the physical limitations and the other's the psychological limitations. So um [name] couldn't go anywhere where she wasn't familiar she would get lost she'd panic, uh so everything she does is within a familiar framework, where she goes, where she walks yeh keeping in that routine and we're lucky to live in [name of place] cos it's quite flat and there is reasonably good walks around the boating lake and so on that are quite safe and straight forward so it's possible for her to do"*

Symptoms causing unstable gait and balance, leading to falls were cause for concern across the FGs for caregivers and people with HD from early stage onwards. Later in this key theme, the consequences of the 'fear' of falling are reported. The possible physical consequences of falls impacting on PA for somebody with HD is illustrated here by a caregiver (spouse) of a woman with early to mid-stage HD in FG7:

*"[...] walking I hold her hand 99 percent of the time just in case she has a little stumble or a slip because she's not as quick, as I said, myself and if she ends up banging her knee or damaging herself she won't be able to exercise and I don't want to take the risk...especially when we're outside on concrete. Around the house it's not so bad. But outside or different areas that she's not used to"*

A number of people with HD and caregivers across the FGs identified impaired coordination of movement as something that made it difficult to do certain activities. For some, this difficulty caused too much of an obstacle because of the extra concentration required, and affected their participation. This is not entirely surprising as difficulty with attention to tasks that require increased concentration are seen to emerge as part of the cognitive impairment in HD (Thompson et al. 2010). PA that requires increased concentration maybe more challenging for somebody with HD and could be a barrier to their engagement or continuation in the activity.

Person with HD, FG6:

*"...I am so badly coordinated like the things like aerobics or classes or whatever, I just can't do it, I'm the sort of person that if I can't do things I don't want to know."*

Some people with HD described experiencing fatigue. The fatigue seemed to have consequences for PA but did not necessarily stop them from engaging in PA as has been previously reported for people with MS (Kayes et al. 2011; Smith et al. 2011).

Indeed, linked with key theme 3 encompassing 'strategies', there was an understanding that fatigue needed to be managed in relation to PA. For example, participants (caregivers and people with HD) talked about planning their routine so that they would avoid planning to do PA at the time of day when they knew that they would be tired.

Caregiver (spouse) FG4:

*[name] has to think in terms of capability. And that, she never chooses lightly. She mentioned already to you about feeling tired, at about 3 o'clock she basically goes to bed because she's practically exhausted by that time in the day. Like lots of things, exercise isn't really an option, becomes a decreasing option. Possibly in the morning she'll walk the dogs then in the afternoon your priority is really about rest rather than exercise."*

Person with early stage HD, FG4:

*"I do try and do jobs and things in the morning er, like, I do jobs and then sit on the sofa. I do all the moving and the cabinet and stuff like that. But yeah, it's just like you know, random tiredness that hits me sometimes."*

Fatigue was also discussed as an issue by caregivers of a woman with mid to late stage HD in FG2 from a different perspective. HD may impact on PA behaviour, but PA could also affect the fatigue experienced in HD. This further supports the idea that if and how fatigue affects them may be something for people with HD to be aware of when planning PA:

*[...] if she gets fatigued it means everything, it effects your balance and it has a knock on effect two or three days after. It's just trying to find that, the right balance.*

Another concern that may have implications for PA was the weight loss that some participants with HD experienced. Weight loss has been seen as something that occurs as HD progresses. For those in the healthy population weight loss might be seen as a positive result of engaging in regular PA, and a motivation for doing PA. However, in the context of HD, too much weight loss may not necessarily be healthy and considering diet and nutritional calorific intake when engaging in regular PA may be helpful. A caregiver clearly explained the importance to their spouse of eating regularly to maintain their weight. This was also reflected in other participant experiences:

Caregiver (spouse) FG7:

*"It's [exercise] probably number two to food isn't it? Food's very important so at the moment she eats five times a day at regular intervals because, erm well,*

*because she doesn't want to lose weight as you probably know people with HD tend to lose weight as they burn the calories up really quickly. So again that's really important to her. She does get a little bit erm, agitated and stressed if she doesn't get her food at the right time."*

Cognitive symptoms were described strongly as being the main challenge for one of the caregivers of a spouse with HD in FG4. The cognitive symptoms appeared to be more disabling in terms of confusion and affecting confidence in comparison to the physical symptoms. This may speak to the pathological changes found in the brain prior to manifest motor symptoms by Tabrizi et al. (2013) and is reflective of the difficulty with complex thinking tasks that have previously been described in the early stages of HD (Kirkwood et al. 2001; Rosenblatt et al. 2000).

Caregiver (spouse), FG4:

*"The thing with [name] is the disease is primarily a cognitive thing. There's the twitching and so on but the biggest feature is the confusion. You know she finds it very difficult, she was finding it difficult to think about the question and it's that cognitive impact that's the big issue on her confidence. It's not a physical incapability to exercise, more a problem actually being aware of enough to go out and do that stuff and remain normal in that respect."*

Another participant with early stage HD in the same FG (4) also shared his experience of cognitive challenges:

*"It takes us longer to process what people are saying to us and then takes us longer to process the answers."*

Both HD participants in FG4 were at the earlier stages. Even at early stage, this comment highlights the need for others to be aware of how it takes some people longer to process and respond to information. This could have implications for PA participation, in terms of receiving and responding to instructions. In turn this could affect time spent doing PA overall because if there are lots of instructions to follow it will take longer. Examples of when this could be relevant include taking and following instructions in an exercise class, gym, sports coaching, following an exercise DVD in the home or instructions for functional activities. Issues of communication in HD have been highlighted previously in a more general context (Hartelius et al. 2010). Following interviews with people with HD, Hartelius et al. (2010) found that for successful communication people with HD required conversation partners to understand the potential difficulties that mean questions or instructions need to be short and focussed.

A caregiver in FG3 highlights this issue:

Caregiver, FG3:

*"Oh and having the patience with her. That's something a lot of people don't have...the patience for her to do things independently."*

The potential for compromising the independent PA of a person with HD is portrayed in the above quote. Lack of understanding that extra time is needed for people with HD to process information, instructions and thoughts (Walker 2007) may lead to impatience. Impatience may lead to interfering, and taking over, and not allowing the person with HD to maintain their independence.

It was obvious from the discussions within the FGs, that for some people with HD, the margins are blurred when it comes to the manifestation of symptoms. i.e. some behaviour may relate to behavioural issues whilst other behaviour could be a product of cognitive deficit. An example of this is given in the summary below by a caregiver. The caregiver felt that it was important for the person supporting their spouse in PA to understand the nuances of HD, how it may affect the behaviour of the person they were working with and how to deal with such circumstances.

Caregiver (spouse) FG7:

*"When [name] met his personal trainer, I sat with them and had a meeting with [name] and her and I explained about Huntington's disease. I gave her leaflets I got from the HDA. I also explained how different it can be for each individual and explained [name]'s symptoms and his needs and whatever. And the main thing that I felt I needed to explain to her initially was the mood differences. Because sometimes he's in a great mood, sometimes he's not. Other times he may not respond when you ask something or speak to him but it's not because he's being rude, it's because of the condition and explained how everything closes down for him and that basically the best thing to do is to give him time and things like that."*

Lack of motivation was identified as an issue for a range of participants with HD across most of the FGs; also recognised by the caregivers. Behavioural symptoms were also identified as barriers to PA participation, in addition to the physical and cognitive symptoms already mentioned. This is consistent with the existing limited literature pertaining to experiences of PA in HD where lack of motivation was found to be a common barrier to exercise across the studies (Zinzi et al. 2009; Quinn et al. 2010; Khalil et al. 2012; Frich et al. 2014). As previously mentioned, apathy has been recognised as an early symptom of HD (Naarding et al. 2009; Krishnamoorthy &

Craufurd 2011) and is a result of pathologic changes which may explain why some people at early stages had difficulty motivating themselves.

Person with early stage HD, FG2:

*[...] one of my problems as I said, I'm lazy, I can't be bothered hum... I find it very difficult to sort of have hum a routine at least for me it would be very difficult to have sort of a routine time for exercise. I got to be honest, I sleep...a lot, I have great difficulty in sort of getting up in the mornings, getting up, my wife shouting at me this morning you know, cos very often I can stay in bed until 10 or 11 and because I've become slower at sort of washing and shaving in the morning, it takes me a good hour just to get ready to do or go anywhere and ah, I've found I've become lazy, I can't be bothered to do these things and I think for me to turn around and say to have ah right you're going for a walk today...it would be difficult, for me anyway"*

Linked to the subtheme of 'Interactions' in key theme 2, verbal tics or 'vocal outbursts' were described by a caregiver as being a worrying aspect of HD whilst exercising in public. As such this could be perceived as a barrier to particular environments when participating in PA. This has been reported for the first time here in the context of HD and PA.

Caregiver (spouse), FG7:

*"Another consideration is...it's one of [name]'s particular things, is to say inappropriate things to people. So if he's in a gym session and there's beefy fellas there, he'll say inappropriate things to them. So there's fear of getting a smack in the mouth as well [...] 'See look at him, he looks like he's on steroids!' See things like that."*

A poignant reflection is given by a gene positive/prodromal stage person (FG8) that indicates some of the turmoil that a person could experience when receiving a positive genetic test for HD. It seems that this individual has made positive steps forward through engaging in PA (indeed he was in training for a marathon). This positive behaviour following receiving a diagnosis reflects findings of Hagberg et al. (2010). Semi-structured individual interviews were used by Hagberg et al. (2010) to explore the physical, social and emotional impact for participants knowing that they carried the HD gene following a genetic test 5-14 years prior. Participants shared their perspectives that new uncertainties arose such as uncertainty about symptom onset and manifestation of HD but also described trying to do enjoyable things while still healthy. In talking about being gene positive, the young man in the quote below draws attention to the risk of depression and what that could mean for PA behaviour even at the very early prodromal stage.

Depression has been recognised as characteristic of this stage in HD (prodromal/gene positive) (Kirkwood et al. 2001; Rosenblatt et al. 2000). Positive predictors of depression include illness perceptions of identity and perceiving the cause to be related to chance (Arran et al. 2014). Arran et al. (2014) suggest that having goals may be a way of achieving a sense of meaning and 'control' which is one of the illness representations of the SRM. If perceptions of control are modifiable then one could argue that targeting negative perceptions could have a positive effect on depression. This could be significant in terms of HD progress as Marder et al. (2000) describe that more rapid functional decline was found to be associated with depressive symptomology in their investigation of rate of functional decline (Marder et al. 2000). The participant underlines his thoughts on the feeling of helplessness that knowing one has HD causes. He explains his experience and that he thinks people may go one way or another. In terms of either thinking that it is pointless to do anything (regarding PA) because the decline is inevitable, or think positively to try and take control, referring to his fellow FG participant who talks about "fighting back" and dispelling the 'black cloud' through exercising on the treadmill.

*"Your head...I think when you find out you've got HD or you've got a gene or you're carrying it I think your head...you can let it go one of two ways. You can either let it drag you down which prevents you from doing exercise and you've got to get out of that negativity or you can run with it and try and do what [name of other person in FG] is doing and try to use it as a good thing and he's really letting his energy go you know...a lot of people I think mentally put obstacles in the way when it comes down to it. Because you feel useless I guess and you go sort of in to yourself and introverted you think...sort of...you think about yourself all the time and you become more at home. So you put obstacles in front of your exercise. You've got to overcome those as well when you find out that news."*

In addition to the identified functional consequences in terms of activities of daily living that people faced, the symptoms also caused worries in relation to PA. The fear of the person with HD falling was common for participants across the spectrum of HD, and caregivers were more vocal in this issue.

Caregiver (spouse), FG4:

*"I think there is a concern now uh about now for [name] about tripping and falling we've been out a few times and she's just gone straight over the edge of the path or something and fallen over so I do really a lot of the concentration is on that."*

The caregiver in the quote below (who is also a Specialist Huntington's disease advisor; SHDA) is obviously speaking from their personal experience which does not mean that it applies to everybody. It does however contribute to the picture that is being built up about the challenges that people might face in engaging in PA. It is possible that the successful story of the woman in the FG doing exercises in the home resonated with this caregiver in thinking about his experiences. It seems he has reflected on this story which led him to make a suggestion to the FG. The suggestion was that exercise in the home could be a preferable alternative to going elsewhere for organised PA considering the behavioural challenges he speaks of:

Caregiver, FG1:

*"I think it's probably a fair observation that organise is a challenge for us to get people out of the house at a specific time on a specific day with the right gear umm and that's a challenge, it's not impossible by any means and there must be a proportion of people who would go for that. But it's quite a challenge, if the objective is to get people to exercise I would guess you're more likely to have success if you could get people to do some sort of activity in their own homes at a time when it suited them, because it might be that they simply want to do it an hour earlier or an hour later...that's perfectly feasible if you're doing exercise at home but if you've arranged a specific time and a place an a venue to do communal activity then I think it's quite a big headache to make it happen reliably."*

Caregivers shared the challenges that they faced when trying to support people with HD to be active. For example, in FG2, the spouse of a woman with mid to late stage HD commented:

*"It's also a syndrome that you probably come across and all carers have come across, they'd rather listen to somebody else than listen to you. You know what I mean. If, if you said something to [name], she'd listen to you. I could say the same thing and it's 'don't keep on, don't keep on'".*

This comment was met with agreement from other members of the FG who had cared for relatives with HD and was seen in other FGs too. The challenge may simply be the behavioural nuances of HD that caregivers face or could be something more complex to do with the dynamics of the relationship. Over time, it is reasonable to consider that behaviour may become more challenging. However, it does not seem to be inevitable from looking across the FGs and instances where challenging behaviour towards the caregiver (and or family member) is mentioned. Where it is mentioned, the participants with HD tend to be early to mid-stage onwards rather than earlier. This may be a reflection of diminishing independence with time and disease progression, meaning that caregiver input is needed, combined with wanting to retain independence and not wanting to take directions or advice from others.

Insight was shared by caregivers regarding accumulation of symptoms over time causing barriers to build up that make it difficult to continue with PA. A caregiver quote from FG6 illustrates this perception succinctly:

*“...as you get deeper into the disease you are unable, physically unable, and probably mentally as well hum to exercise. I think with all the goodwill in the world, you know, they’d love to do it, but they are unable as they get further into it.”*

Of note in terms of the contributions to the FG discussions, an explanation by a person with HD would sometimes be followed by further explanation from the caregiver. As observers of the behaviour of the people they cared for, they were clearly well placed to comment, and in most cases there was agreement between the caregivers and people with HD. If there was disagreement it was generally the caregiver correcting the person with HD. For example, in FG8:

Person with HD: “*I cycle everyday*”

Caregiver: “*You used to cycle every day. You don’t do that now*”

The quote above also serves to remind the reader of the underlying presence of ‘time’ and specifically ‘change over time’ as a thread throughout this theme. It was recognised from the FGs that what is perceived to be PA alters with progression of the disease, barriers change over time, and reasons and motivations change over time too.

In summary of this key theme, the underpinning and salient issues and perceptions across the FGs relate to the changes attributable to HD in terms of abilities, expectations and motivations to engage in PA. Chapter 5.7 discusses the aspects of this key theme in relation to the other key themes in more detail. However, in summary of this key theme, the name given to it; ‘the evolving representations of HD and physical activity’ encompasses the idea that over time, as with progression and changes caused by the disease, so too do the illness representations of HD that individuals develop as a result of the physical and emotional experience of it in relation to PA. Indeed, the illness representation constructs described by the SRM (identity, control, coping, cause, consequences) are applicable to understanding the impact of the nuances and complexities of HD on PA. Perceptions of the disease regarding the physical and psychological symptoms or ‘consequences’ and the impact that HD has on personal identity were elicited as important influences on PA participation. In relation to the research questions, the experiences of PA participation in HD have been elicited across the stages and how some of the nuances of HD influence engagement in PA, which are explored further in the following key themes.

## **5.5. Key theme 2: The varying social environment of the person with Huntington's disease and the impact on physical activity**

The social environment of the person with HD was emphasised across the FGs by caregivers and those with HD as being important in influencing engagement and participation in PA. The social environment of the participants encompasses the immediate physical surroundings, social relationships, and other services and institutions with which they interact. Within the overarching key theme relationship constructs between the individuals with HD and their caregivers (formal or family members) are highlighted. The support provided by caregivers that enabled people with HD to engage in PA was portrayed as integral to the lives of a large number of participants, and with consensus across the FG.

Linked to support provided by caregivers and a common thread across the data was also the burden that can accompany caring for and supporting somebody with a long term illness. Social interactions whilst engaging in PA were highlighted as important, whether for negative or positive reasons. Identity in terms of social identity and role identities (which links to key theme 1) within the family unit were salient issues for a small number of participants in relation to PA as well as the stigma of having a visible disease. The physical environment emerged as an influencing factor of PA in terms of barriers, facilitators and perceptions of different environments.

The sub-themes comprising this key theme are; **preference of environment (in relation to PA); the evolving role of the caregiver in HD; stigma of a visible, genetic disease; support in different guises, and; the nature of interactions whilst being physically active.**

This key theme highlights the vital role of the caregiver in supporting the individual with HD to continue PA participation when the disease progresses and abilities change. This key theme gives insight into the nature of self-regulation in response to cognitive representations of PA capabilities due to HD, linking to the process described by the SRM. The findings suggest that caregiver input becomes more important with progression of the disease to support and help the individual adjust to what is realistic for them regarding PA. In essence, with disease progression, the self-regulation of activity becomes a more collaborative regulation of activity between the caregiver and individual with HD.

### **Preference of environment**

The concept of physical environment in relation to PA in HD refers to the environments in which people with HD liked to be active. It also relates to experiences where the physical environment was a facilitator or barrier to PA. Various environments where people with HD were active included attending group exercise classes or the gym, hydrotherapy sessions, going outdoors, exercising in the home and in social clubs. Others considered functional activities around the home to be part of their PA routine.

Person with HD, FG1:

*“...cos I like to be outside enjoying the exercise to get fresh air ... it’s quite fun being off road. And it’s much nicer if it’s sunny. It has been rather hard the winter all the snow and everything.”*

For some, the physical environment they chose was influenced by their enjoyment of the social aspect of PA. Some preferred to join exercise groups with other people with disabilities and for the social aspect of group exercise. Similar findings have been reported in MS (Learmonth et al. 2013) where people reported enjoyment of an exercise class environment with other people with MS.

Person with HD, FG1:

*“[In a group] You kind of feel you’re not the only one with it, people are worse off.”*

*“...as a social occasion as well it’s very important...”*

Although in MS, group exercise has been found to be favourable, there were mixed feelings for people with HD and their caregivers about group exercise. They referred to the heterogeneous nature of HD where even if at the same stage, people could have different needs and abilities cognitively/physically/behaviourally. The heterogeneous nature of HD can be attributed to differences in the length of the genetic mutation and environmental factors environmental modifiers. Modifiers such as social environment and lifestyle are also thought to be influential but are still being explored (Georgiou et al. 1999; Anca et al. 2004; van Dellen and Hannan 2004).

The outdoor environment was considered to be a barrier by some caregivers who were spouses of people that they described as having unsteady gait and balance deficits. This was linked to the fear of people with HD falling; a fear held by both people with HD and caregivers.

Caregiver, FG2:

*“...because pavements are pavements, they are not perfect...”*

Person with HD, FG3:

*“I’d be worried about falling over.”*

Poor or good weather also influenced outdoor PA for some people.

Caregiver, FG1:

*“I think that as the weather’s got worse and the cold and the snowing and the rain. It, it is quite dangerous to try and manoeuvre [name] outdoors.”*

Person with early stage HD who enjoyed cycling regularly, FG1:

*“...it’s much nicer if it’s sunny.”*

### **The evolving role of the caregiver in HD**

Differences emerged from early stage to later stages in the FGs that highlight the evolving role of the caregivers over the course of HD in light of PA engagement. The nature of caregiver burden appears to change with their evolving roles in supporting the person with HD. This reflects findings from previous work that identified the changing nature and meaning of roles within the family unit in HD (Williams et al. 2009; Røthing et al. 2014). Across the FGs, caregiver burden was identified from discussion of caregivers of those from early stage onwards before development of overt physical symptoms. The changing nature of the caregiver role highlighted the different needs of people with varying symptoms at different stages of HD. Caregivers highlighted and described different ways they provided support for PA participation. The roles ranged from providing simple prompts, to educating personal trainers about HD, providing transportation and actually doing PA with the person with HD. Complexity of HD is highlighted by the discussions that even in earlier stages, cognitive impairment means caregiver support can be required. This finding is in accordance with previous findings in HD (Williams et al. 2007). Williams et al. (2007) conducted FGs with family members of people who were gene positive and at prodromal stage. They found that dynamics within the spousal relationship begin to change as they notice subtle alterations in cognition, behaviour and functioning.

At earlier stages, the caregiver role appears to be less hands on (generally verbal reminders/prompts) than at later stages when the symptoms are more physically disabling. In a phenomenological study of quality of life in HD, caregivers described

how their role is made difficult because the disease ‘keeps evolving and changing’ (Aubeeluck et al. 2012). The changing nature of the caregiver role in HD also appears to be relevant in the context of PA.

For those at prodromal stage and had attended with their spouse, the topic of ‘caregiver role’ did not emerge as a salient issue. However, a gene positive participant shared their insight into how they thought this would change. This is not surprising as he disclosed that his father had late stage HD and so had a clear idea of what could be ahead:

*“...obviously they would have to cope with my caring and she does that very well but she says she has to work at understanding what’s going on and supporting us all emotionally.”*

Caregiver burden related to PA of the person with HD recurred across all of the FGs with the exception of FGs three and five. The caregivers in FG3 were all formal, paid caregivers. In FG5, the participants were residents of an assisted living facility. There was only one informal caregiver in FG5 who was the mother of one of the participants that lived in the facility. Being paid as a job to care for non-relatives is potentially quite different from living with and caring informally for a spouse or child twenty-four hours a day, seven days a week. This could be why caregiver burden did not emerge as a salient issue in FGs three and five in which the caregivers were all employed. An older man in his 70’s with early stage HD summarised his perspective which captures the essence of this subtheme:

*“I think with Huntington’s Disease, I think it’s a two-person illness rather than, or maybe more than two, hum, you know, it’s the person whose diagnosed and it’s the people involved with them; wives, husbands, children hum... all just as involved with the disease as we are you know, the people who suffer...”*

For the informal caregivers, who were mostly spouses, caregiver burden was clearly a salient issue even at the early stages due to cognitive difficulties rather than physical impairment and was discussed 1) more generally with regard to daily activities, and 2) regarding burden related to supporting PA.

Caregiver burden in HD has previously been identified to have a cost to the health and quality of life of caregivers (Williams et al. 2009; Aubeeluck et al. 2012; Cox 2012). Caregivers spoke plainly in the FGs about the burden that they experience. In addition, psychosocial consequences of the strain of daily, and indeed long term responsibility of care in HD, due to isolation and stigmatism of HD have been previously confirmed (Røthing et al. 2014). The burden described by the caregivers and associated

consequences found in previous studies is entirely understandable given the complex losses of motor and cognitive function, and psychiatric symptoms. For example, a caregiver (spouse of female with early stage HD) shares this insight:

*"For a carer it's about having a situation where the person you're caring for doesn't have independence which robs me of mine..."*

Caregivers are amongst the most influential people in the social network of the person with HD, and could support PA participation (Kaptein et al. 2007). It is possible then that caregiver burden may negatively affect engagement in PA for people with HD if the caregiver themselves is personally struggling to cope. If the caregiver is finding daily life difficult in supporting their family member/spouse, then this may impact on their ability or energy to support PA, whether or not it is a priority for them. A caregiver gave insight into their life in a poignant comment where the stress of caring and the importance of PA to them to help them to cope was disclosed:

Spouse of woman with early stage HD, FG4:

*"[...] part of the motivation for me [to exercise] is the heavy drinking, to cope with the stress, cos I drink a hell of a lot so running is a kind of trying to stay healthy in that way drink cos facing reality."*

The perception of burden in supporting PA emerged from a number of perspectives. One caregiver describes the feeling of almost being abandoned to support the person with HD to exercise when they did not feel they had the expertise. Spouse of male with early to mid-stage HD, FG7:

*"They taught me what exercises to do with [name] in the pool...but the trouble is I've forgotten half of those... You need somebody professional in there really. You do need a professional. I don't know do I? And nobody else is in the pool with us."*

Another caregiver of a woman at mid to late stage described how they did not want the added 'pressure' of supporting their spouse to be active within their already full caring routine:

Caregiver (spouse) FG2:

*"Whereas if you took her to the pool yourself, you'd have to be looking after her all the time and arranging to get her dressed... you actually want someone to come along and help you and do that for you, something else that involves you when you're already working 24 hours a day caring for someone and then you've*

*got to take them swimming and get involved, that's another thing that's putting pressure on you."*

Also common amongst caregivers who were spouses of people at mid to later stages across the FGs was the responsibility that they felt for the safety of the person with HD when performing PA.

Caregiver, FG6:

*"Yeah, I worry about the risk with all of it because he does fall hum... and I understand what he's saying he was assessed for a wheelchair but he's refused to have one because he thinks if he doesn't use it he will lose it, so... I've just got to manage the risk."*

Although caregiver burden has been somewhat explored in the HD literature, the observations of the caregivers are reported here for the first time in terms of the burden they perceive in supporting PA. This may be of significance for health care professionals to be aware of when developing activity plans for people with HD. It might be helpful to consider in what ways they can support the caregiver to support the patient so as to relieve the burden. Another example of caregiver burden that emerged from the FGs was lack of time for the caregiver as illustrated by this participant in FG7:

Caregiver, FG7:

*"Even if I go out walking with [name of spouse with HD] you walk for so long. Then I've got to push him in his wheelchair which is blimin hard work... If I could get out on the bike occasionally it would be nice. I used to ride my bike everywhere and now I don't at all. I don't get any time."*

It is clear from the quote above that some caregivers in the FGs perceived their time to be limited because of caring for somebody with HD and emerged as being a barrier to their own PA participation. Previous research has also found lack of time to themselves to be a recurring issue for caregivers of people with HD. In addition this can negatively affect caregiver quality of life (Williams et al. 2009; Aubeeluck et al. 2012; Cox 2012). Specifically, in PD, supporting spouses to be active can have an emotional and physical cost to the caregiver (Jones et al. 2008). In contrast, if their spouses could be, or were supported by somebody else to be active this was considered to be positive by caregivers in the FGs as they could have time to themselves:

Caregiver (spouse) of woman with mid to late stage HD, FG2:

*"...she went for a number of sessions in the [hospital] for hydrotherapy, and she absolutely loved it... I would lead her into the pool and I would sit there, I'd take*

*my newspaper with me and I'd stare for 15 minutes and just read the paper, you know, and knowing she's enjoying what's happening."*

As the caregiver role evolves and they become more involved in the daily activities of the person with HD, the decisions made in terms of what PA is feasible following appraisal of experiences seem to become more collaborative. This is reasonable considering that the shared experiences when caregivers are supporting PA may lead to joint decision making by the caregiver and person with HD. In relation to the process of self-regulation described by the SRM, the FG findings suggest that regulation of PA behaviour in the context of HD becomes more collaboratively regulated over time with caregiver input.

### **Stigma of a visible, genetic disease**

Across the FGs caregivers and people with HD indicated that more awareness and better understanding of HD in the general public and amongst health and care services is needed. Exasperation and weariness of having to deal with other people's misconceptions was obvious through terms they used such as 'distressed' and 'sick of the way' they were treated. One of the caregivers summed up their frustration succinctly with:

*"We never asked for this illness you know"*

Perceptions of being stigmatised because of disability caused by HD and the symptoms of HD were expressed across the FGs. This was portrayed to have negative connotations for people wanting to engage in PA and is evidenced throughout the presentation of this sub-theme. One example is that of a difficult situation explained by a caregiver where his wife was still able to walk some distance but had the visible altered gait and balance problems. She was still trying to be independent but was rejected because of how she appeared:

*"she felt she could walk enough so she could go to the bus stop and catch a bus in, she had her bus pass [...] she couldn't drive, but then she had a altercation with a bus driver, he thought that she was drunk on drugs or something like that [...] eventually her walk became such that she couldn't really risk going on her own on the bus"*

Participants who felt this way and described such experiences included those at earlier stages engaging in higher level activities (such as exercising in a gym) and those performing more functional activities at later stages (such as walking around the shops). Various negative experiences when being active were attributed to how people

with HD are perceived by the public and the public's reactions. Hirsch et al. (2011) suggest the need for strategies to address anxiety about being active in public for people with PD because of the fear of being publicly humiliated. Indeed, embarrassment and worry about how the general public perceive them has been identified as being a psychosocial barrier to participation for people with MS and PD (Elsworth et al. 2011; Learmonth et al. 2013; Smith et al. 2011). The same authors report embarrassment as being the most common barrier to participation for participants of their research who had PD. This is also relevant to HD given the example below where a caregiver gives an example of when she and the woman with HD that she cares for go out to the shops as part of her plan to keep active. The caregiver reassures the woman with HD who worries about what other people think of her.

Caregiver of woman with late stage HD, FG3:

*[She's worried] what other people think, hurry up let's go, like getting in and out of the car, she's like uuuuhhhh I'm taking too much time, I'm like, don't worry, okay, it takes as long as it takes! [...] So there is a stigma attached to, you know, I'm disabled, I have to [...] get out of the way sort of thing"*

A specialist HD advisor of the HDA described the negative psychological effect of public perception she has seen when working with families (FG1).

*"...the anxiety of going out on their own and people thinking that they're drunk so that just limits people then when their partners come out with them and they're having to hold on to them..."*

An example of personal embarrassment for the person with HD was given by this caregiver/spouse in FG8:

*"...there became a time when he was too embarrassed to go to the gym because he felt people were looking at him. And you know, they were! I had to go and support him... in the gym when he was doing his weights because of balance and co-ordination and things..."*

This caregiver (FG7) explains the frustration in dealing with other people's comments and perceptions:

*"People say what's the matter with him...oh he's got Huntington's disease? Well I've never heard of that. [...]A lot of people haven't. That's the trouble. They don't understand. In fact 9 times out of ten they look at you as if you're drunk and that annoys me."*

The stigma mentioned in the quote above sounds similar to how people with MS have previously described their negative experiences in terms of a distrust of legitimacy of symptoms. Skår et al. (2014) conducted FGs with participants of a physiotherapy rehabilitation study for people with MS. The authors found that in MS, worry about others accepting legitimacy of the disease is attributed to the invisibility of some of the symptoms (Skår et al. 2014). The experiences of the FG participants suggest that legitimacy of the disease is questioned because of the visible symptoms that cause people to appear inebriated. Examples were given where members of the public assume that they are people of undesirable character, social deviants with a discredited identity. Social stigma has also been reported as a barrier to exercise in MS where people have reported being stared at in an exercise setting. Negative attitudes of health and leisure professionals and lack of understanding or knowledge of the condition were also identified (Learmonth et al. 2013).

A caregiver shares their experiences of how the lack of awareness of HD means having to repeatedly explain the condition. The person quoted below is the husband of a woman with early stage HD; a young couple in their 30's with a young family. For a young family coming to terms with the manifestation of more visible symptoms it seems the HD is having a social impact. Frustration is expressed of seeing how people react to hearing of a well-publicised condition compared to HD which is considered a rare disease (Evans et al. 2013) and receives very little media attention.

*"I just feel it's important for people to know you know on an everyday level that there's some issue here because she's starting to act in a way that most people would consider strange. [...] it's a really debilitating isolating disease I mean it's one of the worst things about it. When she got diagnosed she got diagnosed stage 1 Huntington's and then 3 months later she got diagnosed with breast cancer. When you talk to people about cancer 'really sorry' you you know and there's a real sense of solidarity, with Huntington's people go 'what's that all about?' you know and it's incredibly isolating not least cos people don't understand it but also because the characteristics are uncomfortable for people to deal with so they tend to say 'oh that's [name] she's a bit odd' you know."*

Although it was quite distinct that the stigma of HD was a negative factor for people when exercising, an interesting deviant case of a woman at early stage HD suggests the opposite. She suggests that participating in exercise has the potential to enable the person with HD to feel equal to the others exercising in a group although they know that they are different. Later in the disease process, exercising in a group setting with 'healthy' people may serve to highlight differences in ability even more so.

Person with early stage HD, FG4:

*"I mean I think it is you feel like you gonna be like different to everyone else and if you can do exercise it does put you on the same level as everyone else in the group as well."*

The isolation caused by being stigmatised (Goffman 1963) certainly appears to be something that is relevant to the people who participated in the FGs and has been found in other neurological disease such as MS (Skår et al. 2014). In most cases where the concept of stigma arises, it appears to adversely affect people's engagement with PA. Using the SRM to underpin the analytical coding framework facilitated exploration of the social landscape of PA for people with HD because according to the model, social experiences influence illness representations and so it was important when analysing the data to consider these social aspects of PA.

### **Support in different guises**

The topic of support was a common thread across the FGs. Perhaps due to the different 'make-up' of the groups in terms of what stage people were at, 'support' emerged in different guises. In this respect, as an overview looking across the experiences shared in the different FGs, the nature of support for people with HD to participate in PA changes, or needs to change over time as HD progresses. The code 'support' within the analytical coding framework was identified through open coding. In refining the themes, it was clear that the idea of 'support' interlinked with the 'consequences of HD symptoms'. It is apparent from the FGs that more support is required with disease progression for numerous reasons, including the consequences of symptoms in terms of the actual symptoms themselves i.e. cognitive, behavioural and physical. In addition, the knock on effect of the symptoms in terms of fears (fear of falling for example), lack of confidence in abilities (self-efficacy) and accessing environments such as gyms and exercise classes, leisure clubs or walking groups emerged.

Those who provided the support to participants with HD included personal exercise trainers accessed through social workers and the national exercise referral scheme, caregivers (formal and informal) and physiotherapists. Knowledge of HD was described as key by caregivers and people with HD for providing appropriate support to be active. Understanding the problems of the individual has been identified as vital to finding optimal support strategies for PA in PD (Jones et al. 2008). Agreement with this concept is seen from the perspectives of participants in the FGs. They emphasise the importance of knowledge of HD in supporting PA, and describe the consequences of not having that. For example:

Person with HD, FG8:

*[...] the person who I saw initially wasn't qualified to deal with some of the symptoms so I found somebody else who was very helpful...all the issues...balance...and walking, so I wouldn't put a strain on my body and I thought the physio was really good. I had to wait until somebody who actually knew...which is a problem really. People outside of our community don't really know what to do"*

Caregiver, (formal) FG5:

*"I think you gotta understand Huntington's in order to you know to get the exercise whatever right, you gotta understand their condition and how their brain works and how they're processing what you're saying so like if you plant seed night before and they've got that to look forward to it might work [...] so I think you've got to understand about the illness before you know the right way to go about it cos it's such so complex the way they take in information you know and also the way they'll agree to things that doesn't mean they wanna do it. Cos yet it's the first thing they say, [name] is just an example of that, she said 'yeh' when you asked does she exercise, she said 'yeh' and she doesn't."*

Attitudes towards the support of people with HD at the assisted living facility in FG5 is suggestive of the positive approach integral to maintaining independence as much as possible. The caregivers described how they encouraged the participants with HD to feel that where they lived was their home not just somewhere to exist because the HD had led to it. The caregiver explains below:

Caregiver 1 (P5):

*"...so if he miss something and someone gets up and about at like 2 or 3 go they're gonna to the kitchen and order something to eat, so we just you know we cater to what they want to do and it's their home..."*

Support in terms of physically being there to ensure safety when using equipment, as well as physically taking people to attend sessions was commonly described by caregivers. For example:

Caregiver (spouse), FG7:

*"I had to go and support him in the gym when he was doing his weights because of balance and co-ordination and things"*

Particularly for people at more advanced stages of HD, it appeared that support from caregivers was vital to continued PA participation. The SRM takes into consideration

that individuals are influenced by their surroundings including the people around them, which these findings reflect. In some cases, caregivers from the FGs were able to purposefully attempt to influence people's perceptions of their abilities by encouraging them and reinforcing positive praise for achievements. This may in turn influence the illness representations of the person with HD and their appraisal of what they are capable of. For example, emotional support was provided by a formal caregiver in FG3 because of lack of confidence and participant's belief that they could achieve their goals of PA:

Caregiver FG3:

*"I find [name] needs a lot of, umm, positive praise for every achievement that she does because you fear that you're doing it wrong. In, in everything and I don't know if that is part of the illness. [name] questions herself, her ability to do things."*

In contrast to this, a number of caregivers from different FGs who were family members observed that they were not always listened to and at times faced antagonism from the person with HD. This may reflect the different manifestations of behavioural symptoms and overall heterogeneous nature of HD. Some caregivers describe a situation where they are the last person who the person that they care for will listen to when trying to offer advice or encouragement. For example:

Caregiver, (parent) FG1:

*"they go in and get him up in the morning "come on" and he goes there like a little gem and you just think... "Why doesn't he do that for me?!""*

Another caregiver explained a similar situation but spoke more empathically and did not take the anger towards him as a personal insult, rather, he saw it as being difficult for his spouse to accept help because it signified a loss of independence.

Caregiver, FG2:

*"I think [name] gets annoyed when I try and support her doing things, she wants to be an independent person for as long as possible."*

A caregiver (spouse) shared her experience of how her husband had been able to continue going to his local golf course because the club and a few of the members supported him, and took him around the course. This is a good example of how a

positive social environment can have a positive impact for the person with HD even at the later stages.

The caregiver goes onto say:

Caregiver, (spouse) FG2:

*“...the people he played with were just so supportive and the family who run the golf course were really understanding and so he had a really good run for his money...you know he done things a lot longer than most people would probably be able to”*

In the quote below, a formally employed caregiver in FG3 illustrates the benefits of having somebody who is determined to support functional activity:

*“...to get her motivated and actually to get her out, she was quite happy just to be sat just looking at the same four walls and to me, she was just surviving, she wasn’t living. And I think that’s what started it... I kept saying to her; you’re a wife, you’re a mother, you’re a woman. You need to envelope those things and become part of the family again because her husband and everybody was doing the shopping and everything for you. And now [name] does the weekly shop herself, umm, she goes out, she collects her prescriptions, everything that she needs that now has made her become a wife and a mother...again, which she wasn’t having before, when we first started.”*

The findings here report for the first time in the context of HD, perceptions of the different guises of support for PA across the stages. There is a dearth of literature for comparison in the HD context, however there is some relevant literature surrounding other long term neurodegenerative conditions. Social support from others with the same condition was a key facilitator of positive PA behaviour in weekly exercise groups and inpatient rehabilitation for people with MS (Learmonth et al. 2013; Skår et al. 2014). Seeing others with the same condition successfully participating in PA gave participants encouragement that they could do it too. It also provided the opportunity for participants to expand their social network through new friendships which also motivated them. In addition, social support is helpful for people with PD in engaging in an exercise programme (Eriksson et al. 2013). The emphasis on the findings by Eriksson et al. (2013) was on the concept of ‘modelling’ one’s behaviour through the observation of other people’s behaviour, (a key aspect of social cognitive theory). In contrast, the idea of support through group PA with other people with HD did not emerge as a salient theme in the FGs (either positively or negatively). This could be perhaps because there are no group exercise classes or PA groups specifically for

people with HD in Wales or England. Indeed, a lack of such groups was discussed by FG2 participants.

The general idea of PA being an opportunity for meeting other people was identified by a range of participants in the different FGs. This (perhaps in similarity to MS) draws attention to the lack of social contact that people experience in HD. In PD, caregiver support for walking has been identified as essential for monitoring and prompting whilst walking (correcting posture and stepping). Familiarity with the problems of the person they cared for allowed caregivers to use optimal strategies to support their walking. For the most part, caregivers seemed to be successful in supporting the person with HD. The same could not be said for how people perceived provision for exercise or PA within the health service.

Inconsistency in HD of provision for exercise / PA prescription from the healthcare system was highlighted through discussion around services and support from particular healthcare professionals. For example:

Caregiver (spouse), FG2:

*“... I think, we should ask professional physiotherapists maybe to be studying how can we counteract the physical movements [...] yes, the carer can be there they can go through a routine once or twice a day that you need to do it, but the professionals need to find that sort of exercise which probably may not exist at the moment to counter for this sort of thing...”*

Across FGs issues were discussed that pertained to the concepts of ‘accessing’ and ‘receiving’ health and social care. These issues included when help is needed to support the physical abilities of people with HD versus when input actually happens. Also described was difficulty accessing services through healthcare in terms of restrictive processes or restrictions of services and lack of specialist support. Not knowing how to access services that could support PA for the person with HD was also discussed.

Across a number of FGs, caregivers and people with HD stressed that in their opinion earlier input from physiotherapists in particular would be more beneficial:

Caregiver (spouse), FG6:

*“I certainly think, you know as a physiotherapist said to me it’s core strength (group talk) so if you have it earlier (group talk) then you can build up your core strength would have helped. I think as [name] said you need it early on where maybe the nurse or the other people roundabout don’t notice it as much*

*because they think they're still active, but that's when it's actually needed. I mean, they tend to get a physiotherapist involved when it's later on things are difficult."*

Caregiver (spouse), FG2

*"[...] if you've just been diagnosed at 40 years of age and your expected to live to possibly 70 plus then if they can develop those muscles that stop them from doing these things then it could be an improvement for the next 20 years of their lives."*

Nevertheless, for a woman in FG2, input from a physiotherapist at mid-late stage HD led to a profound impact in terms of her physical functioning and quality of life. The physiotherapist worked with the woman with HD and the caregivers. The caregivers in turn continued to work with the woman with HD to achieve improved ability to carry out every day functional tasks.

Caregiver, FG2:

*"... she hasn't been able to manoeuvre the stairs because she's been, more or less wheelchair bound [...] And now we don't use the wheelchair because we've implemented the Physio programme and [name]'s mobility has come leaps and bounds."*

The SHDA in FG1 who worked closely with HD families shared their perspective of the difficulty for people with HD to access services that might support PA:

SHDA, FG1:

*"It's interesting we do the clinics and one thing most families would like is physiotherapy but the physiotherapy treatment around the country is sooo different. You get good treatment in one area and there's absolutely no physios in another. You know they do these falls clinics, so families are expected to go to the falls clinic absorb, retain and remember all of the information they've been given and then say goodbye![...] So you can say this is part of the personal care plan and the direct payments they're a bit more generous with the time, so you can incorporate it [support for PA] into there, cos actually it's essential that they have this exercise cos it's for their well-being and overall safety, building up the muscles and so psychologically, emotionally, psychiatrically, mood wise it's all gonna help. So it's how you approach and if you've got a good social worker who understands Huntington's, then you're there."*

Something that came across strongly because of the shared experiences amongst those in different FGs was concern for the lack of specialist support for HD. The caregiver quoted below highlights this issue, which has been previously identified in HD (Skirton et al. 2010) but not in the context of HD and PA. Where support was received

(for PA through the health service) people were left feeling that they were going around in circles because of a lack of a specific route to appropriate specialised support. This in itself could be a barrier to PA for the person with HD.

Caregiver (spouse) FG7:

*“...you were going to physio weren’t you? With [the physio] but you were only allowed to go to 5 sessions with her and then you had to be referred back to the GP and then you had to wait again and then you could go back to her again”*

Caregiver, FG4:

*“For me it’s ... the fact that when it comes down to it the support for exercise with Huntington’s disease is actually quite specialist and the general problem and the lack of specialist support and general infrastructure is er being gyms and doctors or whatever, there’s no specialist support there to kind of help deal with the specifics of what happens.”*

The caregivers below considered themselves ‘lucky’ to have been able to gain support through healthcare services, suggesting that such support may not be common. This is consistent with previous findings where caregivers resign themselves to the fact that they must become accustomed to not receiving the professional support required (Aubeeluck et al. 2012). Others seemed to stumble across the way in which they received appropriate support as depicted in the second quote below.

Caregiver (spouse), FG7:

*“We were lucky enough to be given the NHS health budget and [name] goes twice a week. She also has a massage once a week which really helps. You have a massage too don’t you?  
[spouse] “Yeah, I love it.”*

Caregiver (spouse), FG7:

*[...]it was only though being assessed for so many hours through the day centre that they were able to change it over [...] unless you’ve got a social worker involved and an assessment going on, you’re not going to necessarily know that can happen because we didn’t know about that beforehand.”*

Barriers for PA in MS and PD (other than consequences of the disease) including lack of services for and signposting by physicians of PA, meaning a lack of opportunities have also been identified (Ravenek & Schneider 2009; Learmonth et al. 2013).

Although for the most part the lack of appropriate services was perceived to be a road

block, one pragmatic caregiver took another approach. The caregiver explained how they had linked in with their spouse's social worker to facilitate access to appropriate individualised support for PA. As well as highlighting the potential importance of the social worker role within the health system to facilitate support for PA in HD, this example links in with the SRM, but in relation to the actions of the caregiver. It appears that the caregiver made an objective appraisal of a situation and took action in a way they thought best to be able to improve the situation for their spouse in terms of continuing PA.

Caregiver, FG8:

*"They assessed [name] for the day centre and that's how we got it. Through direct payments with social services. When he went to the day centre he didn't like it and he wouldn't go, yeah. It wasn't for him is what he felt. And our social worker at the time said 'well, what would he like to do?' and the thing that came up really was the gym."*

#### **The nature of interactions whilst being active**

The pertinent quote below illustrates the perceived importance of PA participation for the social interaction.

Caregiver (spouse) FG4:

*"it's not just about the exercise it's about the solidarity you find in that because it's something that's missing in the lives of Huntington's sufferers."*

Unfortunately for people with HD though, although there were positive experiences of social interactions while being active, instances of negative experiences were not uncommon. Negative social interactions and worrying about having negative social experiences whilst engaging in PA were described by participants with HD and caregivers. These were linked to the more overt symptoms of HD and included the visual gait disturbances, balance impairments and verbal tics (vocal outbursts). The negative social interactions were largely with people of the general public who did not know the person with HD. This links with the subtheme of 'stigma'. i.e. stigma of socially abnormal behaviour or somebody who's appearance deviates from what is considered 'normal' can cause negative social interactions when participating in PA for someone with HD.

Caregiver, FG7:

*"So if he's in a gym session and there's beefy fella's there, he'll say inappropriate things to them. So there's fear of getting a smack in the mouth as well."*

Caregiver, FG2:

*"...she felt she could walk enough so she could go to the bus stop and catch a bus [...] but then she had an altercation with a bus driver, he thought that she was drunk on drugs or something like that."*

Positive social interactions whilst engaging in PA were depicted as friendly competition, praise for achievements from caregivers, interactions with others in classes. Other included people in a walking group ensuring that the person with HD could keep up and support from friends at social clubs.

Caregiver, FG2:

*"we had a very understanding walking group as well who would always try, although if we were ever anywhere a bit precarious I had to make sure they didn't help and it was always me who took the risk, but we really struggled along gamely right until he went into care and it was just our salvation."*

A sample of an interaction between 2 people with HD who take part in the activities on offer at the assisted living facility, FG5:

*"HD1: She went to the bowling one time a couple of weekends ago, [name] beat me the first week and I beat her the next week.*

*Facilitator: Is it good to have a bit of competition?*

*HD1: Yeh*

*HD2: [Giggles] Yeh!*

*Facilitator: Yeh, do you two compete?*

*HD1 and HD2 together: Yeh*

*HD2: I'd win him. The night we went bowling...[pause] I won him, we were bowling weren't we..."*

Another common finding was people with HD using PA as an opportunity to mix and socialise with other people, even to gain social acceptance. Seeking social acceptance may link to the negative reactions from people of the general public that was described in the FGs and discussed in the subtheme around stigma.

Person with HD FG4:

*[reasons for exercising] ... interaction and things like that. The social side, I suppose. Erm, yes, being able to talk to people, y'know having a little chat to people .... Yeah, I think that's important as well [...] I think it's good to um be social and be with other people, interacting as well as you can you know? To a point where they stop noticing things."*

Caregiver FG4:

*"I think for [name] it's a big feature of why she exercises, walks the dog, goes to pilates because it's one of the few areas left in her life where she can actually interact with people outside of that which don't really happen. Social life, going out on a boat and meeting people on the boating lake, meeting people that way... incredibly isolating disease."*

Exercising with other people in a group who also have disabilities and also exercise groups specifically for HD was discussed in FG1, FG2, and FG5. Caregivers questioned whether such groups existed and there were mixed thoughts in weighing up whether it would be helpful. For example:

Caregiver FG1:

*"...have a specific date and a day and a time for people with disabilities not just HD to go along and... for the day, you know they have an hour once a week sort of thing for people with disabilities I don't know whether that's segregating them and then discriminating even more I don't know..."*

Caregiver FG2:

*"...you need specific groups that are tolerant of the type of people we're talking about Huntington's and other, and other.... people who have got walking difficulties etcetera, it's not just Huntington's you know that, but you can't just try a ramblers group who just happen to be going for a Sunday 4 mile walk you know, it's got to be more on the lines of associating with people with disabilities..."*

When an individual with HD in FG1 was asked whether it would bother them to see other people with disabilities when they were doing PA they responded with:

*"Oh no, no not for me anyway. You kind of feel you're not the only one with it..."*

This quote echoes the sense of community elicited from FGs with people who had participated in MS only group exercise rehabilitation (Skår et al. 2014). The study aimed to gain better understanding of how people with MS experience inpatient

rehabilitation and the potential psychosocial benefits. Participants described how being with similar others facilitated sharing of experiences, strategies for coping and adjusting mutual support. They became what they considered to be "colleagues" (Skår et al. 2014).

Being able to interact socially played a key part for one participant's PA in FG5 and different participants had different ideas about what PA is. This diversity of perception is highlighted by a participant of FG5 who considered dancing at a social club (supervised by caregivers) as part of her PA. The physiotherapist that supports the residents of the home she lived in put a different spin in explaining the PA that this particular individual did:

*"...it's more of a social activity with physical activity involved with it"*

In summary, this key theme describes how the social environment in particular plays a key role in PA participation in HD. Discussed in more detail in chapter 5.7, components of the SRM were found to be relevant to this theme including 'coping', 'evaluation' and 'illness representations'. With regards to the main research aim, this key theme highlighted issues specific to HD in relation to the social environment (for example social stigma, social support and interactions during PA), and how they impact on the experience of PA participation.

## **5.6. Key theme 3: Achieving physical activity participation while coping with the nuances of Huntington's disease**

The third key theme very much links to the concepts of the other themes because coping is influenced by environmental factors, previous experiences, understanding and perception of HD and PA. Coping responses are described by the SRM as a key part of the self-regulation process. Individuals are said to perceive a 'health threat' (in this context, perhaps a change in the HD symptoms) and respond with a given action which is then appraised for its success. Examples of this type of cognitive processing are a common thread throughout this key theme, suggesting further relevance of the SRM as a whole. Other research has considered particular components of the SRM, i.e. illness representations (Clare & Harman 2006). To gain the full picture of the utility of a model and its relevance to the topic explored, it needs to be considered as a whole. The relevance of illness representations and the social and environmental influences of cognitive representations described by the SRM have been elicited in key themes one and two. In this key theme, the process describing coping responses is relevant. The overarching concept of coping in the context of HD and PA was identified in a number of ways. These included using PA as a way of coping; trying to maintain normality despite living with an unpredictable progressive disease. Also how coping or non-coping in relation to HD might affect PA behaviour and management of PA through using specific strategies with success.

Participants portrayed appraisal or monitoring of the success or failure of their efforts in terms of their PA and representations of the experiences of HD. This gave invaluable insight into the barriers specific to HD and how barriers evolve over time as people experience difficulties with different issues. For example, at early stage barriers have been elicited as stigma when gait and balance become affected and the person no longer has a normal gait pattern or are able to stand still (woman with early stage HD, FG4). For an individual at later stage the increased gait impairment was found to be such that it formed a barrier to continuing to be active (woman with HD, FG2). Even more helpful in contributing to the overall picture was the insight provided as to how some such barriers were overcome by people with HD and their caregivers.

Pertinent to this theme are the adaptations and strategies elicited from the people with HD, caregivers and healthcare professionals. A highly relevant point is made by a person with HD (below). Already demonstrated by the subtheme of 'the evolving role of the caregiver', this person reiterates that it is not just the people with HD who are coping with the effects of the disease but the caregivers and family members too.

Person with early stage HD, FG2:

*"I think with Huntington's Disease, I think it's a 2 person illness rather than, or maybe more than two, hum, you know, it's the person whose diagnosed and it's the people involved with them; wives, husbands children hum, you people who work connected with it, I mean you're all just as involved with the disease as we are you know, the people who suffer..."*

The subthemes comprising this key theme are: **continually adapting and adjusting expectations of ability over time and tried and tested: strategies for engagement specific to HD across the stages.**

#### **Continually adapting and adjusting expectations of ability over time**

It was recognised across the FGs that different people have different perceptions of what PA is for them, depending on their abilities at the time. The change over time with progression of HD appeared to be pertinent to what people with HD perceived PA to be for them. Those at early and early to mid-stage were more engaged in more energetic exercise at the gym, running, exercise classes, swimming and cycling, gardening either independently or with some support (for example one to one supervision in the gym, transport or support via a GP referral scheme).

Person with early stage HD, FG8:

*"So I do treadmill and also the rowing. They've got a running machine and a cross trainer so I've finally managed to do all of this sort of stuff quite recently in the last six months or so I've started doing it through my err...it's like a GP referral scheme."*

In comparison, for those at later stages of HD who were less independent, PA encompassed more functional type activities. These included walking, home based exercises and classes such as hydrotherapy. For example:

Caregiver (spouse), FG1:

*[...]so I'm sitting there watching TV and [name] is just sitting up getting up and down with this paper cup and this ball [...] a paper cup with a little ball on top that [name] had to keep, now she stood up and sat down without dropping the ball and then like smaller medicine ball..."*

Caregiver, FG2:

*[...] our goal was to push the shopping trolley round Tescos while she's walking. And she achieved that, even grabbing the groceries and putting them in herself."*

For some, the term ‘exercise’ had negative connotations. Participants discussed how PA might be spoken about differently so as not to put people off engaging in it. The idea of promoting, or framing the concept differently of what being ‘active’ is, so that different types of PA are considered and people do not feel pressured to do certain types of PA came across from a number of FGs. In the quote below, the physiotherapist specifically used this as a strategy to facilitate engagement with the activities available to residents of the assisted living facility.

Physiotherapist, FG5:

*“Um but I think with the general population when you say ‘exercise’ to general population I would rather you know use sometimes physical activity or ‘let’s do activity’ or something because when you tell them about for engaging the whole ‘exercise’ seems to be like ‘oh I’m gonna get tired’, oh it’s gonna be like long and strenuous’ ‘oh I’m gonna be asked to do things I don’t wanna do’. So I try to term it now physical activity or let’s do an activity.”*

Caregiver, FG5:

*“...you think about something specific...the gym, but actually people exercise all the time you know it’s just about being out and about and moving about. It doesn’t have to be a specific thing.”*

As HD progressed over time people changed to lower intensity and less cognitively and physically demanding activities that allowed them to maintain some sort of activity. As a physiotherapist, the researcher recognises that with degenerative diseases, the approach taken might change to maintaining function and quality of life, and preventing secondary complications rather than improvement as such. This is usually through encouragement and education about more functional activities such as sit to stand practice, mobility, and balance tasks that include functional elements such as reaching. Such activities are important to be able to continue with activities of daily living such as getting up and down from seats, toilet, and bed, reaching into cupboards for food, or clothes walking safely around the home or outside. Maintaining function to meet physical, emotional and psychological needs contributes to an individual’s well-being despite their declining health. In the FGs, for those who wanted to continue it seemed that they maintained some sort of PA to a degree through adapting what they did in line with their abilities. This suggests that with disease progression there is an adaptive process whereby cognitive representation of the illness experience might guide action planning or ‘coping’ responses and performance of these. This is then followed by appraisal or monitoring of the success or failure of coping efforts. This resonates with the findings of Eriksson et al. (2013) where the process of redefining oneself was been

found to be an underpinning concept with regard to exercise in light of impairments and activity limitations in MS.

For those who have more severe HD illness representations, perhaps ‘exercise’ as an achievable concept is unrealistic. They may see ‘exercise’ as something requiring attributes that they believe they no longer possess; a good level of fitness, good coordination of movement, balance and normal gait. People who spoke about PA using the term ‘exercise’ tended to speak more about activities such as running, outdoor cycling, gym aerobic exercise classes. More people at earlier stages talked about PA that they were involved in as ‘exercise’. Although people at later stages still talked about ‘exercise’ it was more related to their past, previous to manifestation of HD symptoms.

It is apparent from the FGs, perceptions of what constitutes PA differed between earlier and later stages of HD. The SRM helps understanding of this in that it suggests that an individual’s cognitive representation of their illness is partly constructed through their experiences of the illness (Levanthal et al. 1984). The cognitive representation of their illness held by an individual with HD may affect perceptions of what they are able to do in terms of PA because of how they perceive HD to affect their abilities. In turn this may affect what they perceive PA to be and so a difference is seen in perceptions of what PA encompasses for people from earlier to later stage. Somebody with pronounced balance and gait impairment may find walking as challenging as somebody at prodromal or early stage finds running or cycling. Indeed, measures of disability have previously been found to correlate with life goals in people with neurological conditions (Nair & Wade 2003).

It is suggested that as people with neurological conditions adjust to loss of independence in certain activities (including leisure activities), they attach less significance to them as ‘life goals’. Perhaps this reflects a coping strategy to avoid disappointment in not being able to achieve them. This is consistent with findings that in PD, PA goals were dependent on impairments and grade of activity limitations (Eriksson et al. 2013). This appears to mirror the findings reported here somewhat. For example, the woman in FG2 with mid to late stage HD who the caregiver refers to in the quote above. Her PA consisted of working towards realistic, functional goals that were important to her everyday life. In comparison, somebody else who recognised their limitations in speed and coordination but was more able-bodied still felt able to join in a gym class but swapped from an aerobics class to a slower, less intensive exercise class.

Person with early to mid-stage HD, FG8:

*"I found aerobic things too fast. I just couldn't cope with the speed mentally or physically so then they said well, this balance one is a lot slower but still does similar types of things so I went to that. And although I've still got trouble with my co-ordination I'm doing some of the exercises and I'm enjoying it."*

The descriptions given by the woman in FG2 and man in FG8 suggest acceptance or realisation of their capabilities and place more importance on realistically achievable activities or goals that reflect their abilities in the context of HD. The task specific training for a study in subjects with mid-stage HD focussed on addressing functional exercises. These were activities of daily living that were identified as being important to individual participants (Quinn et al. 2014). Using 'Goal Attainment Scaling', goals were set collaboratively between the exercise trainers and participants. The goals were individualised, and participants considered them to be relevant to them. Subsequently 46% of participants achieving goals that were considered 'much better than expected outcome' i.e. they surpassed their original goals, and adherence to the exercise programme was 96.9%.

The findings of the FGs contribute to the description that began to emerge from that study; that people with HD engage more with activities that they consider relevant and achievable. The FGs provide the additional experiential insight that what is considered (and actually is) achievable changes as the disease advances. The social benefits that were gained from adapting were also highlighted. Poignantly for one woman, adapting meant being able to enjoy time out with her husband again but adapting was difficult for her to accept. Although her husband had to persuade her to use a wheelchair for longer journeys, she eventually understood that by using a wheelchair she would not be so tired and achy. Also that she could enjoy being out and doing some walking instead of struggling with long walks.

Caregiver (spouse) of person with mid to late stage HD, FG2

*"I think [name] gets annoyed when I try and support her doing things, she wants to be an independent person for as long as possible...in the balance and all that, she doesn't say it but it must be exhausting for her, but you know, but she's too stubborn to use a wheelchair (group laughter) we just came back from [location] a week ago, we had a week in [location], you know a week in [location] and I persuaded her to hire a wheelchair while we were out there so that meant that when we left the hotel, we could walk out the front and around, and she was able to still see the views and the both of us sit down and have a Latte and enjoy the sun and it wasn't so much effort to get to those places and get back then, so she was, she was able to enjoy being there without feeling tired and aching and suffering from the walk. So I think she realised certainly it came to wheelchairs they were very useful for her."*

Participants also described having adapted where they go and what they do because of the psychosocial effect of being in an uncomfortable social situation. Appraisal of such a situation was described by a caregiver, where people were staring at the person with HD because of his physical symptoms. This led to them changing what they do; an objective response to avoid a potentially awkward situation related to this person's specific symptoms and the PA environment:

Caregiver (spouse), FG7:

*"Well now he's embarrassed about going to the gym, erm, she comes to the house and she'll work with him in the garden and do circuits. She'll take him to the beach and do body boarding and different things like that."*

The example given above suggests a positive way of dealing with difficulties due to HD. It is also worth considering the negative potential for people doing less than what they are actually capable of because of not being able to find a way around such issues. Reduced self-efficacy and fear avoidance as a result of symptoms, was elicited from participants of an exercise programme in MS (Learmonth et al. 2013). Although self-efficacy has not yet been specifically explored qualitatively in the context of HD and PA, there is some limited evidence to suggest that better self-efficacy promotes adherence to an exercise programme (Khalil et al. 2012). In addition, Busse et al. (2017) found that an exercise programme that incorporates promotion of competence led to improvement in self-efficacy for exercise. This links in with the subtheme that relates to reasons and motivations to participate in PA. In the FGs, lack of self-efficacy was an issue for some people with HD as described by this caregiver:

FG3:

*"In, in everything and I don't know if that is part of the illness. [name] questions herself, her ability to do things."*

Experience of apathy was another nuance of HD that influenced people in terms of adjusting their expectations about PA. This comes across very clearly with the individual quoted below. In their FG the participant described having always been motivated to be active. He had completed marathons, army training, but one can see how the apathy associated with HD is affecting him from the quote below. He is evaluating what is happening and has been happening, and is perhaps trying to adjust his expectation of what he is capable of. Even so, it seems he is losing motivation to do much at all. Although he may well still have been physically capable, the apathy was a barrier to his motivation and also had an impact on his expectations for PA. Perhaps in addition to less physically challenging PA he needed something more achievable in

terms of overcoming the apathy; something less cognitively challenging and not as mentally effortful.

Person with early stage HD, FG8:

*"This is it...I've got loads of ideas but I just don't...it's too easy to sit by the telly isn't it and forget every...it's always a good idea when I come back from taking the dog for a walk and stuff like that...then I just sit down in front of the telly and I just forget ever... I need something more achievable, It's not happening now. I've got two slipped discs so whilst I've been in the army I would always do the crazy stuff like running with boulders and logs and stuff like that like they have to do. So I found it quite easy up until now. This year was the point when they worked out there was something wrong so I'm getting local discharge. But yeah, loads, I love it."*

The concept of adjusting expectations has been elicited through the changing identities of people with HD and the expectations of certain roles and the part that PA can play in that. However, adjusting expectations in terms of what people were capable of was also a strong thread, and ties in with the concept of people adapting what they do. There needs to be a degree of adjusting expectations following recognition of a change in an individual's representation of HD in order for them to adapt. Some participants recognised that what they were doing to keep active was not working for them because of the physical limitations. An example is given by a male with early stage HD in FG8:

*"I do errr, it as a group balance group but it's for the people who are there that have had heart attacks. That kind of thing. It's like a very slow aerobic thing for...the whole sort of balance thing is involved as well and trying to do various things with coordination so erm, it was a bit...when I went to the first one I didn't know what. I found aerobic things too fast. I just couldn't cope with the speed mentally or physically so then they said well, this balance one is a lot slower but still does similar types if things so I went to that. And although I've still got trouble with my co-ordination I'm doing some of the exercises and I'm enjoying it."*

This individual realised that he needed to adjust to do the right kind of exercise for him. With the advice of the exercise trainer and swapping the exercises, this individual highlights an example of evaluation and refinement of their activity as described by the SRM.

As already discussed, for some, the focus of his PA was about keeping fit and achieving goals such as running half marathons. There was also an underlying commonality amongst participants in that they felt PA gave them independence. An example where one woman wanted to be independent in her walking but suffered consequences because of falling was shared by her spouse in FG2. This is the same woman who was persuaded to use a wheelchair on holiday to have a more enjoyable

social experience.

Caregiver (spouse), FG2:

*"Well she tries to be active all the time, [name]'s backside is purple and yellow, she's fallen on the floor twice in the last week, she's absolutely rotten, she's black and blue and yellow and purple, she's like a baboon, I tease her she's got an arse like a baboon"*

There are intimations here of the psychological or emotional struggle of giving up activities that symbolise independence. Walking is such a basic part of everyday life for most people. To have that ability and choice taken away is very restrictive and could intensify the impact of HD (Charmaz 1983). Charmaz (1983) describes the loss of self as a fundamental form of suffering in the chronically ill. Indeed, no longer being able to walk independently and adapting to being somebody that moves around in a wheelchair signifies a major change in self and loss of the former self representation. Alternatively, whilst on holiday, using a wheelchair enabled a more pleasant social experience with her husband. As has been elicited by numerous participants in the FGs, the connotations for adapting due to HD involve more than just the physical, functional aspects.

The feeling of 'normality' has arisen more than once throughout the presentation of the findings and has been elicited in other subthemes such as the fluidity of the HD identity. It seems that emotional reactions to progression of HD can drive PA engagement as a coping strategy to maintain function. Perhaps adjusting expectations of what 'normal' is would help in dealing with the emotional response to HD and PA. However, one might argue that 'normal' for the person with HD cannot be a fixed entity because of the ongoing decline and lack of constancy.

Caregiver (spouse) FG6:

*"...independence it is a big thing, I mean say she's been active for so many years and then having to rely on myself, and all this, you know, it gets to her, why sort of thing, you know sorry I can't help you in all this stuff, you know but as I say, during my illness I say she's been there and helped me, say it's my turn now, in nit hey...but hum, yeah she always kept the house nice and clean, tidy and everything else. So basically she was on her feet all the time, but hum, she's got a chair and she sits in that one or the same when we go out. I don't think she appreciates the fact ... she's got no appreciation. She has trouble accepting, accepting the fact that she can't do...Yeah, as I say, she did fight to stay on her feet. You know, but come the end, you know...it, it was impossible."*

Caregiver (spouse) of woman with mid-late stage HD, FG2:

*"[name]'s got one on the side of the toilet, she's got one for the side of the sink so when she's washing, she's got two by the bath to help her in the bath because*

*she doesn't like a shower and she still insists on a bath, I got the thing to put on the bath for here to sit on for lowering in the bath, so we got all that in the house."*

*"Yeh, yeh or just pop to the shops to get a bit of food to try and do some cooking for the kids later...I mean I do try and get out. Even if it's just a walk to the high street to get some food, but I do try and get out and do some exercise during the day. I mean, it varies, at the moment I've got a 13 year old... as much as you can, I suppose try and keep a bit of a routine for her as well."*

The description in the quote above from FG2 suggests that the person with HD had some insight into the limitations caused by HD. To the researcher this suggests development of a cognitive representation of the HD that is in tune with the actual physical symptoms. To paraphrase Levanthal et al. (1998) who developed the SRM, 'expectations are not at variance with the valid, biological representation of the disease'. Despite being limited because of the HD, this person appears to still try to have realistic expectations about what they *can* do and perhaps demonstrates adjusting to adapting and possibly a degree of acceptance that HD is changing her role identity within the family. In PD, cognitive and emotional representations of the disease are suggested to be important determinants of adaption (Hurt et al. 2012) and coping strategies aimed at accepting the consequences of HD are significantly positively related to wellbeing (Helder et al. 2002a). Linked to the findings, it appears that for people with HD, engaging in PA that is appropriate to their abilities (the cognitive and emotional labelling of HD is in sync with their actual abilities) which differs from what they used to do may be indicative of positive coping strategies.

#### **Tried and tested: strategies for engagement specific to HD across the stages**

A strong thread throughout the FGs including participants across the stages was 'finding what works for the individual' in terms of maintaining their continuing engagement in PA. Strategies used specifically to deal with the nuances of HD when trying to engage or maintain successful engagement in PA were discussed. People who were gene positive or had early stage HD described their own strategies for motivation. For example, an individual who was gene positive described the struggle with his 'mind' in finding the motivation to train for the half marathon he had registered for. His strategy was to set himself achievable goals by increasing distance gradually.

Gene positive person, FG8:

"Well it's really difficult to begin with. I gave myself a mile perhaps the first few weeks then increased the intensity to two miles and do it in stages. And now I'm up to 6 miles with 3 months to go until I hit the ten miles so...obviously your mind prevents you from doing a lot of things, particularly in terms of exercise...so once you've done it then that's a barrier and then you keep going up from there on."

The idea of goal setting was something that caregivers also considered to be helpful for their spouses in the later stages of the disease;

Caregiver (spouse) of woman with mid stage HD, FG1:

*"it's not getting 'em to run marathons or compete in the Olympics it's purely just giving them probably a little bit of focus, and a little bit of strength in their body getting that you know the getting the quality of it right, the exercise..."*

Caregivers were more explicit in describing strategies they used in supporting people later on in the disease or with more overt behavioural and or cognitive symptoms (not necessarily later stages). Understanding HD was identified in key theme 2 as being crucial for caregivers, exercise professionals and health care professionals to be able to provide appropriate support for PA. Here the researcher again emphasises the importance of understanding of HD. Understanding the individual's needs and 'quirks' seemed to allow application of appropriate strategies more likely to be successful in facilitating engagement in PA. One caregiver used a strategy of educating the exercise professional who was working with her husband about HD. She recognised the importance of the exercise professional's understanding of HD for her husband's successful participation:

Caregiver (spouse), FG7:

*"When [name] met his personal trainer, I sat with them and had a meeting with [name] and her and I explained about Huntington's disease. I gave her leaflets I got from the HDA. I also explained how different it can be for each individual and explained [name]'s symptoms and his needs and whatever. And the main thing that I felt I needed to explain to her initially was the mood differences. Because sometimes he's in a great mood, sometimes he's not. Other times he may not respond when you ask something or speak to him but it's not because he's being rude, it's because of the condition and explained how everything closes down for him and that basically the best thing to do is to give him time and things like that. Then I went on to the physical side of it. More for health and safety and for her safety more than [name]'s. Erm, if he's there with a couple of bottles (laughs), keep away! Don't let him stand near a mirror, you know! Kind of things like that. And knowing that, starting the basics with him, she's...continuity of the same person is so vital. Because she's got to know him, build a rapport and started very very basic and realised, actually, I can push him and I can do more, maybe because it's from what he's done before. Erm, and that was what she needed to know. She needed the time to spend for a briefing and then the time to build up the rapport."*

The SRM discusses cognitive representations of the self. Where people with HD are supported to exercise by healthcare or exercise professionals, it is important that those supporting them form accurate representations in their mind of the abilities of the

person with HD. With HD this may be more complex because of the non-visible symptoms such as change in mood. This is described very well by the caregiver (FG7) in the above quote and reflected in the HD literature. To understand and know how to deal with the non-visible symptoms and overt motor problems is imperative for exercise / healthcare professionals and caregivers. Such understanding has been found to be beneficial in PD by Jones et al. (2008), where familiarity with the person and the condition facilitated optimal strategies to support their activity. Such strategies included prompts to correct posture and stepping; encouragement; promoting relaxation in difficult situations and using their own rhythm of walking to pace their partner appropriately (Jones, 2008).

Caregivers shared interesting strategies within the FGs that they used for people who were not always motivated to do their PA routine. For example, one of the caregivers in the FG consisting of the group of people from the assisted living facility shared their strategy of planting a seed for PA a day before the PA class. They explained that mentioning the class the day before meant that the exercise class was in the person's mind sub consciously and so would not be a surprise on the day. People with HD like routine, they can become quite rigid in what they will or want to do on a daily basis but for this person in particular, if the expectation is there, then they are more likely to do it.

Caregiver at assisted living facility, FG5:

*"Sometimes if you plant the seed the night before, can be with, she's out, something's gonna happen like today, that sometimes works, so maybe swimming starts it, and say tomorrow we're going swimming she might spend today thinking about it therefore, get up and go."*

The physiotherapist working at the assisted living facility agrees with the caregiver but also added to this. He highlighted the differences between people and that you cannot always predict an individual's behaviour:

*"I think for all participants here, planning is very important, but for [participant] it's different. He will just want to take swimming there and then and will ask you to go swimming and just want to walk, walk like out of the blue and ask to go walking or like he will be happy to go on the on the bike there and then and you know, a different approach."*

A caregiver from another FG below reiterates this in thinking about the routine she used to organise for her husband before he went into a residential home. Here yet another aspect specific to HD was highlighted; getting stuck on a thought and obsessing over it.

Caregiver (spouse, FG2):

*"he needed to know what he was doing and then there wasn't really any question about it cos that's what we were doing, you know, the only thing....there again is you know if you put Monday go for a walk, well as soon as he's up he's in the porch getting his shoes on to go for a walk you know [laughing] and it's no good going not yet we're not going til this afternoon cos every 10 minutes he'd be out in the porch putting his shoes on to go for a walk, but hum... basically the motivation is if you know you are doing something that day you will more than likely do it."*

The physiotherapist in FG5 shares their use of distraction technique when one of the residents becomes obsessed with something:

Physiotherapist, FG5:

*"Erm we work 9-5 if [participant] in the morning we could schedule um which service users to do try to fit in maybe not the whole hour session probably 20 minutes, or 15 minutes so it stops it... so we'll have other distractions... really just something for it have to get his mind off that whatever he wants."*

In the specific setting of the residential home, PA was something that was regularly on the agenda of activities for residents. The physiotherapist emphasised how he would use 'information giving' about the benefits of exercise generally and for HD as a strategy to motivate people to engage.

Physiotherapist, FG5:

*"So what we try and do for someone who hasn't been like is again um I think sometimes they relate it to Huntington's disease not only exercise is generally good for everyone, makes you feel good, but the exercise is helping the Huntington's disease so maybe we should have exercises regardless of what."*

Similar to this, Eriksson et al. (2013) suggested that an important part of deciding whether to participate in PA was the influence of the healthcare professional advice given concerning exercise. Wanting to gain more understanding of exercises in terms of the potential benefits of strength exercises was also a motivator to exercise following participation in a progressive resistance exercise programme for people with PD (O'Brien et al. 2008).

Another strategy that a caregiver in FG7 used to facilitate walking was taking a wheelchair on a walk so that they could use it if or when needed, and were not limited in being able to go out. This is similar to the story of the couple that went on holiday and used a wheelchair so that the woman was less tired from walking which made the holiday more enjoyable.

*“...we always take the wheelchair with us because he isn’t always going to make it there and back.”*

Another specific strategy that caregivers reported to be helpful in supporting people with HD in their PA was to support their confidence and self-efficacy in activities. Such an approach is supported by the evidence from a study that tailored a PA intervention in HD to individual needs and took an approach that supported autonomy, competence and relatedness. The results indicated that the intervention approach led to improved self-efficacy for exercise and increased levels of physical activity (Busse et al. 2017).

Caregiver of person with mid to late stage HD, FG3:

*“...she was never left in the situation where she would fall. It was always lose her balance but we were always right behind her, even though she was independent, you knew we were there.”*

Formal caregiver, FG3:

*“We have implemented that, we’ve got a communication book now that we’ve put down for her husband and everything she achieves we set her goals and we set a day we’re going to try and achieve something new.”*

It is apparent that it is important for people with HD that others understand the consequences of living with the disease and how that may impact on PA. In addition, understanding within the relationship between the caregiver and individual with HD was perceived to be important. This understanding or knowledge becomes more vital to successful PA participation for the person with HD over time, echoed by the subtheme of ‘support in different guises’ of key theme 2. An example of a caregiver knowing how far they can prompt the person with HD is given in the exchange between a woman with HD and her formal caregiver below. It seems that they are able to reach mutual agreements through negotiation. This allows the woman with HD to still feel she has the ability to choose and make decisions. Rather than being a passive receiver of care the woman is involved in her care but still allows the caregiver to encourage and support her to be active appropriately.

FG3:

Formal caregiver:

*“I’m tired.” Off comes the duvet and she says that she didn’t want to get up—oh sorry, didn’t you? Okay! And then the bed comes up and she’s like I wanted to stay in bed! Oh dear, well we’re up now. Let’s get up!”*

Woman with mid-late stage HD:

*“Haha deafness from you.”*

The physiotherapist in FG5 also refers to the idea of establishing rapport with the person with HD to facilitate engagement in exercise.

*“You have to first I think establish a relationship, you have to gain your trust because you are asking them things to do. So I think it’s important that like you communicate with them and establish first a rapport, a relationship and maybe it might not lead directly to exercise because exercising could be not part of their, or [inaudible] so I’m not gonna do exercise, I like try to talk about their other interests or perhaps like they want to do the rest of the week to choose the programme so I think that that works, for them a new patient, either the music or the other social activities. A typical example [participant] is not really very keen unless he wants something, so there should be an incentive really at the end of the session, so he gets either like I’m taking him out or give him what he wants after an exercise session.”*

Formal caregiver, FG3:

*“With our schedule and our routine that we’ve sort of got ourselves into, so we’re know what we’re doing and we tend to make plans in the week. Well if you do this with me, then you’ll get your lay in on Tuesday and if we then do this on Wednesday then we can manage that on Thursday for you to have an extra hour. But we will be doing this on Friday... Yeah, umm [name] knows that for every thing (pause) it’s it’s difficult because when she’s doesn’t want to get up she’s very tired, umm but there are things that need doing. I’m not there to do it for her.”*

Some participants discussed strategies such as keeping diaries and calendars on the wall as visual prompts as reminders about appointments and exercise sessions at the gym.

Person with early stage HD, FG8:

*“Because I’m medically retired I’ve got more time now so I have actually started doing the gym and I’ve had to put it...because I don’t always remember...put it on the calendar every time I’m supposed to be going and then make sure that I go...so... I’ve found if I have it on my calendar and I sort of almost treat them like appointments so I’ve got to go. If it wasn’t there I wouldn’t do it. I am easily side-tracked.”*

Finding the right routine, maintaining a structure and prioritising according to perceived need for the person with HD was perceived to be important in managing HD in terms of PA. Interestingly this was something that arose in relation to people across the stages. In the context of HD, it seems where integrating PA into daily routine is important because of the cognitive difficulties people even at the early stages experienced with planning, remembering and prioritising. Also for some, prioritising PA was difficult because of the numerous healthcare appointments in addition to other activities of daily

life. For example, the caregiver below talks about how the woman with HD has worked out a routine where her priorities are ensured to be met which include eating at regular intervals and exercising;

Caregiver (spouse), FG7:

*"And then I think on a Tuesday it's about 11.15 until about 12.15. Something like that. So [name], sometimes she'll eat a bit early then so instead of 11 o'clock she might eat at 10.30, so she has something then, give her food a chance to go down and then she'll be exercising just after 11. For an hour at least. And then she'll have her dinner, well lunch, at about 12.30 or quarter to 1... [name]'s priorities are food, exercise and sleep"*

Caregiver (spouse) of person with early stage HD, FG4:

*"For us, the exercise is in the routine. Get up in the morning, especially on a school day we've got a very strict and set regime, times to be, daughter to get to school, breakfast to make, house work to do and [names]’s main exercise comes in that morning period where she'll do housework stuff and then structure in her walk down to the high street to pick up some food for the evening meal and then we've got the dog walk and the rest of the afternoon to rest so there's a kind of a structure to the day, for us, again, the priority is staying in that structure as much as we possibly can. It's all familiar, it's not intellectually challenging it's involving some level of physical activity which can constitute as exercise and it seems to function quite well and I think the longer we can kind of keep it in that place the better off we're going to be...Y'know that's partly why we got the dogs, it makes you go out otherwise...one of the big dangers, unless you've got a structure in your life it can be quite difficult to find that motivation."*

The woman with HD added to this:

*"I try and get out with the dogs or you know go for a walk to like get shopping and stuff like that so I try and do that every day really but I also get tired as well don't I so by the afternoon I've lost all my energy and I'm more likely to be on the sofa so, but if I'm gonna something I try and do things in the morning."*

In addition, the formal caregiver of a person with mid to late stage HD finds routine to be important for the person with HD, FG3:

*"So when you bring the routine into it, [name] has always said that, umm, she doesn't like the routine, the routine doesn't help but I think now she would alter your opinion on that because I think a routine does help her."*

Planning, it seemed, was a vital part of maintaining a regular routine that incorporated the necessary daily activities including PA reflected in the quotes below. The specialist HD advisor of FG1 below gives their interpretation, summarising their viewpoint having worked with a number of HD families. A first-hand account is also given by a caregiver of FG3.

Specialist Huntington's Disease Advisor, FG1:

*"Patients I've worked with I've found fatigue has been a contributing factor to them not going out cos they're just too too too tired and quite often they're hungry as well cos they've forgotten to eat cos actually reminding them to eat before they go anywhere you know get something to eat, families making sure they eat before they go out and they've gone to the toilet that sort of thing [...] it's all about planning yeh, regimented, it's like a military precision isn't it getting it"*

Caregiver, FG3:

*"Making sure we've got the time. 'Cos [name] is very time orientated, with having to collect [her daughter] from school...so you have to be back in time, time is one thing."*

For some, planning involved keeping diaries and writing appointments on calendars kept in plain sight helped people to plan.

Caregiver, FG7:

*"And the only way to keep sane is to keep going, keep it all written down"*

In addition, the need for planning exercise around the timings of taking medication was recognised. A caregiver in FG7 underlined that there can be implications for PA participation because of medication side effects. The caregiver placed emphasis on the need for understanding how medication affects people with HD and consideration of implications of medications for PA.

Caregiver, (spouse) FG7:

*"...if medications changed it can affect the person. When [name]'s was changed a while back he was zombified for a little while, and it's until the meds find their own level...So therefore when he's zombified he doesn't feel like doing exercise. He can't do any exercise. His body's not working. His brain's not working. Once the levels been found in the medication that's been altered or changed he's alright again. There needs to be an understanding of that process as well. That time. That is somebody's meds are changed they're not going to feel the same as they did the week before until everything's working ok and everything has to find its' level again."*

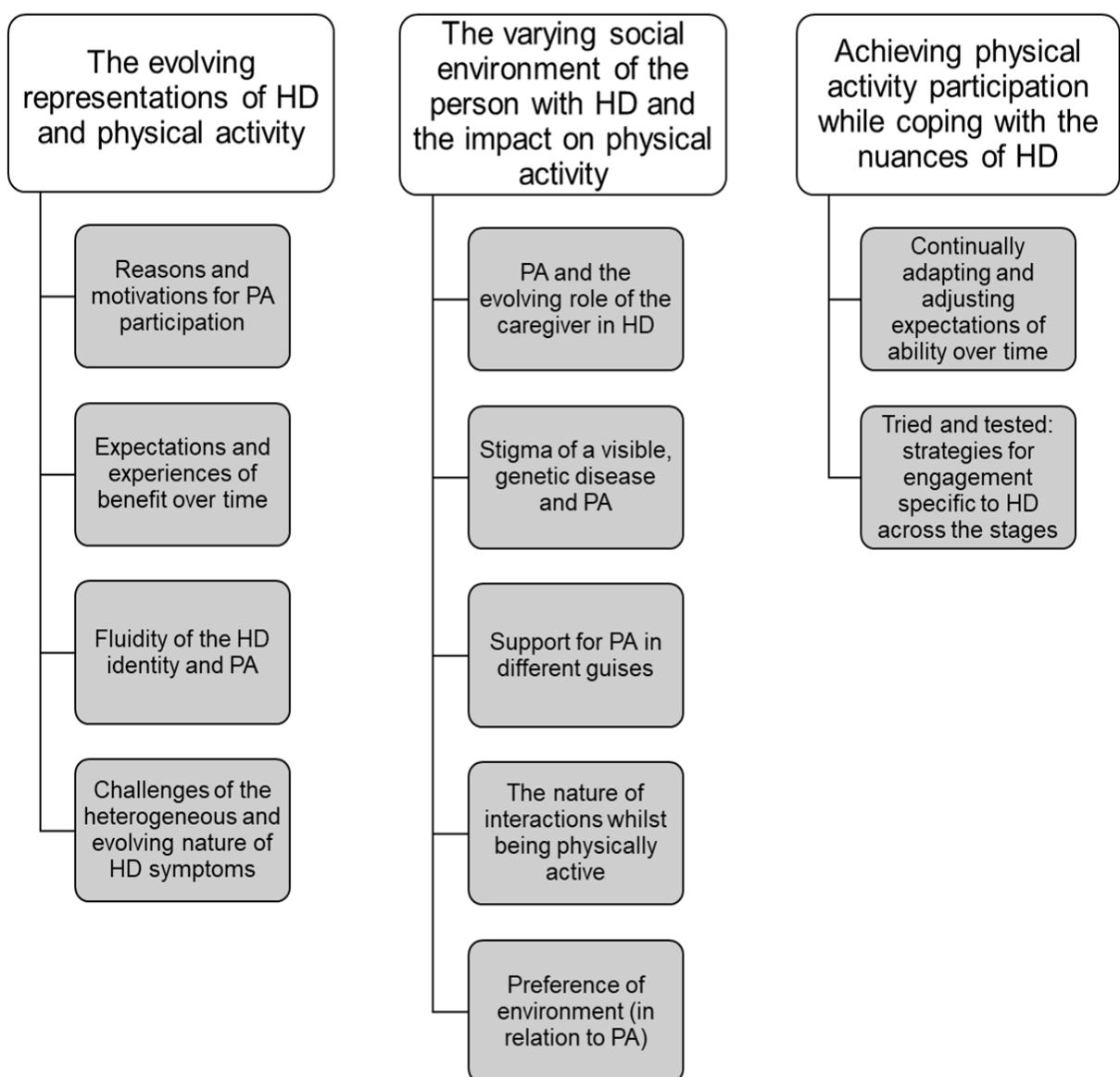
Managing fatigue and fitting exercise around it was described by one couple. In previous exploration of PA experience in MS, managing limits and acknowledging body sensations that indicated fatigue was important for participants (Eriksson et al. 2013). Perceived control over fatigue may influence PA participation positively if managed successfully (Smith et al. 2011).

In summary, this key theme, 'achieving physical activity participation while coping with the nuances of Huntington's disease', encapsulates participants' experiences of adjusting their PA over time to adapt with the progression of the disease, and tried and tested strategies for maintaining PA as abilities change. Finding what works for the individual in terms of maintaining their continuing engagement in PA was key in relation to the continual adjustment of expectations as well as what people do in terms of their PA. The subthemes ('continually adapting and adjusting expectations of ability over time' and; 'tried and tested: strategies for engagement specific to HD across the stages') also discuss relevance of the SRM which is also elaborated on in the following section 'drawing together the key themes'.

## 5.7 Discussion: drawing together the key themes

This research aimed to address the gap in knowledge of the perspectives and personal experiences of physical activity (PA) in HD and contribute to the limited body of evidence regarding PA experiences of people with HD. The key themes presented and discussed in the preceding sections describe experiences of PA participation from the perspective of people with HD and their caregivers, and how the nuances of living with HD affect engagement in PA. This section aims to draw together the key themes of which the reader is reminded in figure 14.

Figure 14: Diagrammatic Representation of key themes and subthemes

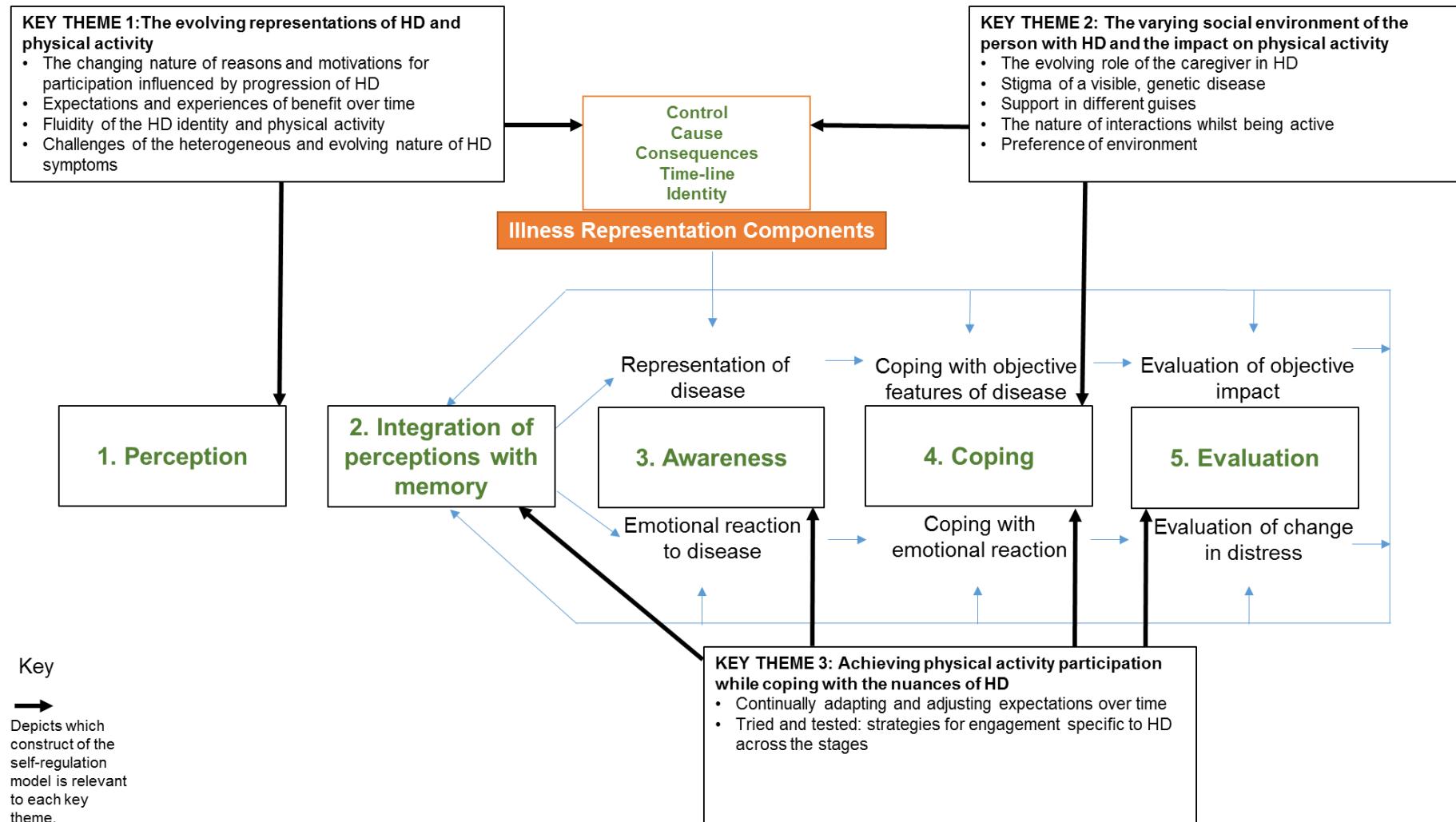


Throughout the presentation of the results there are links between key themes and subthemes; they are distinct yet interconnected and contribute to the overall picture

that has emerged from the research. In particular, in theme 2 there is overlap within some of the subthemes. The researcher did not merge such subthemes because they are important individual concepts and in reality, the environmental influences that form an individual's world do not exist in isolation. Indeed, people live in a 'messy reality' and participants constructed their realities and shared experiences that were embedded within an environment of complex social, environmental and cultural influences (Yilmaz 2007). Examples of overlap are seen in 'the nature of interactions whilst exercising' and 'support in different guises'. Each aspect influences the other thus making it impossible and inappropriate tease apart and consider individual components in complete isolation.

The lack of theoretically underpinned qualitative research in this field led to the identification of the self-regulation model (SRM) as a potentially useful model to facilitate understanding of PA experiences in HD. As discussed within the results, components of the SRM are directly relevant to each of the key themes. This is represented in figure 16 to show this visually. Key theme 1 links to 'perceptions' and 'illness representations', key theme 2 relates to coping and illness representations. Key theme 3 is shown to relate to the lead up to the coping response in terms of 'awareness', the action taken, 'coping' and 'evaluation' which feeds back so that perceptions of consequences of actions are integrated with memory 'integration of perceptions with memory'.

Figure 16: Illustration of the key themes mapped onto the Self-regulation model (Levanthal et al. 1984)



Key theme 1 'the evolving representations of HD and physical activity' encompasses issues that directly affect how HD is perceived by people with HD i.e. the illness representations of HD that individuals develop as a result of the physical and emotional experience of it in relation to PA. Living with HD (whether family member/caregiver or person with HD) gave rise to perceptions of the disease regarding the physical and psychological symptoms or 'consequences'. Those cognitive representations of the disease and the impact that HD has on personal identity are important influences on PA participation. Salient issues and concepts across the FGs relating to the changes attributable to HD in terms of abilities, expectations and motivations to engage in PA underpin key theme 1. The illness representation constructs described by the SRM (identity, control, coping, cause, consequences) were applicable to understanding the impact of the nuances and complexities of HD on PA.

Within key theme 2, ('the varying social environment of the person with HD and the impact on physical activity') experiences of social stigma, social support and interactions during PA are described. The importance of appropriate social environment for PA participation in HD is highlighted in this theme. Experiences of PA are seen to influence the cognitive representations people have of their HD and guide coping responses and appraisal or evaluation of actions in relation to their PA participation. The components of 'coping', 'evaluation' and 'illness representations' within the SRM are relevant to this theme.

Key theme 3 'achieving physical activity participation whilst coping with the nuances of HD' describes how people from prodromal to late stage HD have developed strategies to engage in PA. Encapsulated within this theme are participants' experiences of adjusting their PA over time in order to adapt with the progression of the disease, and tried and tested strategies for maintaining PA as abilities change. It also describes the continual adjustment of expectations as well as what people do in terms of their PA. The components of the SRM that describe adjusting, adapting, developing new representations and integrating those into memory relate to concepts within this key theme.

### **Change over time: Physical activity across the stages of Huntington's disease (prodromal to late stage)**

All three key themes are linked by the overarching concept of change over time, which derives from the progression of HD over time. Time is one of the components that contribute to illness representations described by the SRM. This concept is pertinent to the key themes because change over time from prodromal to later stages of the

disease impacts on how people with HD see themselves (linked to key theme 1), how they engage with and are affected by their social environment (key theme 2) and how they cope (or not) and adapt, in relation to PA (key theme 3). Perhaps the importance of 'change over time' was to be expected in a neurodegenerative disease where there is decline in health over time. Up to this point, participants of this research, who were at different disease stages, had already seen older generation family members decline and or had experienced it themselves. This experience may be different in the future, particularly with the exciting gains in research exploring preventative and disease altering approaches within the last decade. However, translation into practice and achieving engagement in potentially disease-altering activities such as regular PA will be an ongoing challenge. Engagement in activities described in the FGs portrayed the picture that activities perceived to be PA for different individuals was associated with progression of HD over time.

For people at different stages, various experiences of how HD affected them also elicited different approaches to PA. Concepts relating to perceived consequences of HD, issues of identity and being in control influenced for example, whether people engaged in vigorous exercise versus functional physical activities, their reasons for being active, and the challenges they faced. The components that comprise the social environment or 'social landscape' for the individual with HD also changes, because of disease progression. This also has consequences for how people with HD participate in PA, including the social stigma of a visible, not well known disease, interactions with others when being active and support to be active.

Consensus across the FGs contributed to an overall agreement that PA needs to be encompassed across the disease spectrum and that the social environment in particular plays a key role in PA participation. Similarities to previous findings in studies of MS and PD and the small amount of research in HD were found regarding perceived importance of supporting engagement in PA (Dodd et al. 2006; O'Brien et al. 2008; Zinzi et al. 2009; Quinn et al. 2010; Khalil et al. 2012; Eriksson et al. 2013; Learmonth et al. 2013; Skår et al. 2014; Frich et al. 2014). In contrast to the findings by Quinn et al. (2010) and Frich et al. (2014) there was a lack of instances in this research where participants had experienced the opportunity to develop a personal exercise plan with healthcare professionals, perhaps highlighting a gap in services. The geographical spread of the FGs highlighted differences in what was available to people with HD in terms of support for PA through the healthcare system. The differing experiences of people at various stages of HD suggests that approaches to supporting PA from the

healthcare service requires flexibility for people with HD, taking into account their individual needs and abilities.

Across the stages of HD there is clear variation in the barriers that people with HD and their caregivers experience in trying to be physically active or support PA. Quinn et al. (2010) who report some disease specific physical barriers, namely managing gym equipment due to the movement disorder. The research findings presented here build on the work by Quinn et al. (2010) and the FGs elicited various physical, cognitive and behavioural consequences specific to HD that affected participation. Challenges depicted by participants included physical difficulties with walking, balance, coordination, feeling that they were too slow, social barriers, apathy, forgetfulness, inability to plan or prioritise. With symptom progression over time those barriers evidently become increasingly challenging to overcome. It seems that participants perceived the barriers due to the symptoms of HD are not going to 'go away' as such but may be overcome to an extent, linking in with concepts within the key theme 3 'Achieving PA participation while coping with the nuances of HD'. What is perceived to be PA alters with progression of the disease, barriers change over time, and reasons and motivations change over time too.

The concept of 'challenges due to the physical environment' was discussed more by people with HD and caregivers for people at later stages. This might be expected because of the progressive manifestation of physical and cognitive symptoms. Psychological barriers pose a risk of decreased PA but are perhaps also modifiable (Helder et al. 2002a). The fear of falling when being active was identified as one such psychological barrier by both people with HD and the caregivers from early stage onwards. The fear of falling has not been identified as a salient feature in HD, however the only study to have investigated this is limited to a small sample within a quantitative study (Grimbergen et al. 2008). Approaches which may suitably alter perceptions around the concept of falling and risk, and education around falls in HD may be fitting and improve PA participation. The proposal of influencing beliefs in the context of illness representations in HD has previously been supported to improve patient well-being (Helder et al. 2002a). The concept may also have a place in influencing illness representations to positively influence PA behaviour.

Participants gave numerous reasons and motivations for PA participation from both the perspectives of the caregivers and people with HD. A similarity seen in other long term neurodegenerative conditions was the poignant motivation that being active enabled participants to fight the disease and take control in some way (Ravenek & Schneider

2009; Plow et al. 2009). Previous exploration of illness representations suggests that people with HD report little personal control (Arran et al. 2014). Yet there are examples of individuals in the FGs using PA as a way of empowering themselves to feel in control. This may speak to the findings of Arran et al. (2014) where people feel out of control and want to take back control but also gives additional insight into how people try to do this using PA. Motivation to participate in PA was also facilitated by perceived personal benefits.

Similar to findings in other long term neurological conditions, the perceived benefits included better lifestyle and health, quality of life, maintaining independence and functional abilities for as long as possible, and opportunity for social interaction (O'Brien et al. 2008; Kasser & Kosma 2012; Eriksson et al. 2013). The perceived benefits described by FG participants certainly resonate with observations of the researcher and conversations with participants of exercise programmes that the researcher previously supported. In particular, comments about feeling better in themselves and perceived functional improvements were recurring. The process described by participants of engaging PA behaviour, perceiving benefit and thereafter being motivated to continue with the behaviour portrays the idea of self-regulation (Leventhal et al. 1984).

### **Self-regulation of physical activity in Huntington's disease**

The SRM suggests that individuals construct a dynamic cognitive representation of their illness through their own experience, the social and cultural context (Levanthal et al. 1984). The key word it seems for HD is 'dynamic'. In this sense, there is a need for constant self-regulation because of the changing nature of the disease and new representations that form over time. Key theme 3, 'achieving physical activity participation while coping with the nuances of Huntington's disease' encompasses interwoven concepts attributable to the concept of coping. The 'coping' component is a key part of the SRM and is suggested to follow awareness and development of a cognitive representation of a perceived health threat. It appears that for the FG participants this is also relevant in the context of HD and PA.

It appears that the cognitive illness representation of HD may play a role in adjusting expectations of what the individual with HD is capable of doing in terms of PA in adjusting to / coping with the limitations caused by HD. Discussions during the FGs highlighted cognitive linking of social, bodily and emotional experiences of HD to the impact on PA and how participants responded in terms of coping strategies (or lack of ability to cope). As well as giving descriptions and discussing, participants

demonstrated reflexivity at times. Participants reflected during the FGs about strategies that they had used to good effect or not so good effect and how that changed what they did and/or how they did it.

Through sharing their thought processes, participants highlighted that they have engaged in self-regulation. In the context of PA and HD, people have responded to their experiences of PA with certain coping strategies, evaluated them in terms of success, and modified behaviour if conceived to be necessary. Support to self-manage where appropriate is one of the delivery expectations under the 'Living with a neurological condition' theme of the delivery plan 'Together for Health - A Neurological Conditions Delivery Plan' (Welsh Government 2014). The importance of supporting development of skills tailored to individual needs and local situations to encourage self-management has been indicated in stroke (Jones et al. 2016) but there is currently no guidance specific to HD for self-management in relation to PA. Successful self-management (incorporating PA) in neurological conditions is thought to be strongly influenced by self-monitoring and exercise self-efficacy (Dobkin 2016).

Dobkin (2016) argues that cognitive impairment may become a barrier to self-management of PA as impaired self-awareness, memory loss and mood can impact on achievement of self-efficacy for PA. Given the decline in cognition associated with progression of HD, this could impact on PA for people with HD. Examples of lack of ability to concentrate and needing reminders for PA were elicited in the FGs. Cognitive impairments caused by HD over time may indeed limit the capacity that people have for self-regulation. However, following participation in a HD specific exercise intervention which incorporated behavioural support strategies, people with early to mid-stage HD who were cognitively impaired achieved increased self-efficacy for exercise (Busse et al. 2017). Intervention participants in this study had lower scores on cognitive tests than age-matched healthy controls (Tombaugh et al. 1999; Sheridan et al. 2006). Dobkin (2016) argues that cognitive training of planning and multitasking has potential to help achieve self-efficacy for exercise. It might be argued that some element of cognitive training was an outcome of the ENGAGE-HD intervention because it incorporated collaborative repeated planning and reviewing of individuals' programmes over a period of time. Or it could be that the tailored support of the intervention which took into account disease-specific issues including cognitive impairment, helped compensate for cognitive impairments. For example, the weekly reminders via telephone calls to intervention participants may have prevented participants otherwise forgetting to follow their exercise plan.

Other behavioural support strategies used in the intervention were goal setting, personalised feedback and encouragement (Busse et al. 2017). These were strategies also elicited from FG participants. Goal setting in particular was one strategy that was used from prodromal through to late-stage HD and has been used in interventional studies in HD (Quinn et al. 2012; Quinn et al. 2016), but there were also different strategies used by people at different stages such as adjustment of activities, prompting, reminders. In line with previous research of PA experiences in PD, goals were dependent on physical limitations (Eriksson et al. (2013). Quinn et al. (2010) and Frich et al. (2014) found that motivation was positively influenced by understanding of the purpose of exercises, goal setting and participant collaboration in developing a personal exercise programme rather than being given a prescribed routine. Goal setting was certainly a positive feature for some FG participants (particularly a woman in FG3). There are similarities across HD, PD and MS regarding strategies used but these findings further the knowledge of how people living with HD can achieve PA participation in spite of the complexities they face specific to HD. Goal setting and other strategies elicited from the FGs suggest that some participants with HD had taken steps to self-manage through PA, and for others, their caregivers were supporting self-management through supporting PA. Given the potential for maintenance of function, and independent living for longer, this is an encouraging finding.

The SRM suggests that individuals respond to awareness of an issue by problem solving and goal planning leading to an action plan (Levanthal et al. 1984). The action plan may be further refined over time following further appraisal of the success of it. In terms of responding to appraisal and evaluation of PA it seems obvious, perhaps automatic, that if something is not working, common sense leads one to change what they are doing. However, in the context of a neurodegenerative disease it is not so straightforward. For an individual with HD, changing how they do something may signify progression of the disease to them. However, there is evidence to suggest that coping mechanisms related to acceptance of HD have been positively related to well-being (Helder et al. 2002a).

In the context of dementia, people at early stage were found to exhibit self-maintaining responses or self-adjusting responses in the face of progression of the disease (Clare & Harman 2006). The tension between maintaining one's previous identity and adapting to make life easier because of symptom progression was certainly highlighted in the FGs. Within the context of the wider literature, adapting, adjusting expectations and using specific strategies have been elicited regarding PA in MS and PD (Eriksson et al. 2013; Skår et al. 2014). In terms of general strategies in HD, Maxted et al. (2014)

found that in family dyads people used asking for support and humour to deflect from difficult situations. Strategies specific to facilitating engagement in PA were elicited as maintaining a structured routine, encouraging knowledge of others in supporting people with HD to be active, and ways that people with HD motivated themselves.

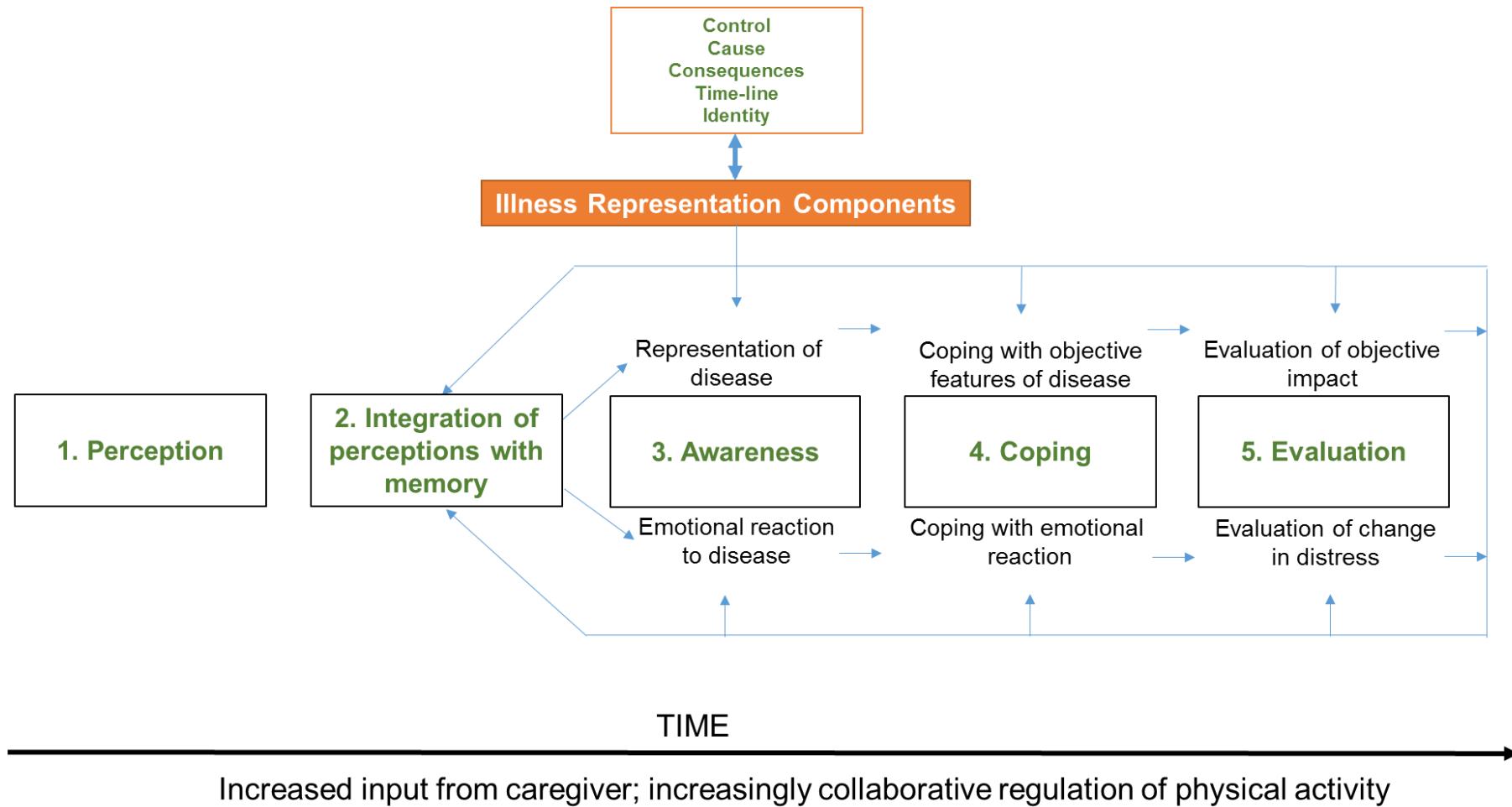
Specific to HD, Khalil et al. (2012) reported strategies to overcome barriers to PA as prompting and cues from caregivers, similar to the findings of Jones et al. (2008) in PD. Two key strategies found by Jones et al. (2008) included monitoring walking using concentration and correcting walking through generating rhythm and size of steps. In the FGs, caregivers' knowledge and understanding of the individual with HD was elicited as key to supporting their PA using strategies that they knew worked for the individual. The dependence on caregiver support in HD has previously been elicited with regard to general personal experience of HD (Williams et al. 2007; Downing et al. 2010; Aubeeluck et al. 2012; Maxted et al. 2014). The findings suggest that this is also relevant in the context of PA. Due to the progressive nature of the disease decreasing capabilities of the individuals with HD meant increased need of input from caregivers.

The findings highlight need for change in support and change in role dynamics between family members over time with HD progression. With these changes, regulation of behaviour becomes more collaborative rather than entirely self-regulated as caregiver input becomes so vital to the daily life of the individual with HD. In other words, regulation of PA behaviour in the context of HD becomes less independent over time. This is seen regarding caregivers supporting people with HD to reconsider and refine what they do in accordance with abilities. Perhaps it is that the caregiver becomes a more prominent source of feedback or information in terms of informing changing representations over time and becomes more integral to the coping responses. The researcher suggests that an increasingly collaborative approach to PA participation is needed over time and therefore suggests an adapted version of the SRM specific to PA participation in HD (see figure 17).

In the adapted model presented by the researcher (figure 17), the cyclical nature of the model has not been altered as the findings suggest that the 'feedback loop' in terms of awareness of changes, regulation and adapting in relation to HD and PA is relevant; highlighted by the key theme 'evolving representations of HD and their impact on PA' and 'achieving PA participation while coping with the nuances of HD'. The element of time is important because over so many cycles of this 'self-regulation' the disease will be progressing. This is why the arrow is along the bottom of the feedback loop, separate from it, because regardless of whatever is happening with 'self-regulation',

time continues to pass and progression of HD continues. As key theme two (the varying social environment of the person with HD and its impact on PA) highlights, the passage of time is directly linked to progression of HD and the need for caregiver input related to PA, and increasingly collaborative regulation of PA is seen. As such, 'time' is depicted with 'increased input from caregiver; increasingly collaborative regulation of physical activity' beneath it to show that this is a consequence of the passage of time.

Figure 17: The proposed adapted self-regulation model specific to physical activity in HD



## 6. Research conclusions and implications

In this concluding chapter, the original contribution to knowledge is presented along with discussion of clinical and research implications. Limitations of the research are also addressed.

### 6.1 Original contribution

This qualitative research provides an original contribution of work that has not previously been addressed, and has explored how living with HD impacts on the experience of physical activity (PA) participation from prodromal to late stage. For the first time, the Self-regulation model (SRM) has been used to facilitate exploration of PA in HD and as a result of this research, a novel, adapted version of the SRM has been developed within the context of HD and PA. Key considerations for research intervention development and physiotherapists and other healthcare professionals working with HD families to support PA have been identified.

The original contribution of new knowledge gained from this research includes:

- The importance of the caregiver in supporting the person with HD to engage and participate in physical activity, which over time and with disease progression becomes increasingly integral. Taking into account the social and familial context of individuals with HD needs to be a key consideration when health care professionals are supporting and encouraging them to participate in physical activity. Strategies such as using PA as a way of taking control, and adjusting expectations can help people with HD continue to be active in different ways and adapt their PA from less high level to more functional activities over time with progression of symptoms.
- The social landscape of the individual with HD impacts on their participation in physical activity. This includes the stigma that people with HD experience when engaging in physical activity in public due to symptoms of HD that can be seen as socially undesirable by those not familiar with the disease.
- The self-regulation model (SRM) facilitates understanding of the impact of HD on PA across the stages of HD from prodromal to late stage in terms of the relevance of the process of regulation that people go through. However as a result of this research, a modified SRM is suggested which takes into account the increasingly collaborative regulation of representations and PA participation with HD progression over time.

Understanding the impact of the nuances of HD on PA participation regarding symptoms, coping responses and strategies, social environment, support needed, changing illness representations and abilities is critically important given the emerging support for PA as an intervention for people with HD. The growing research base that

supports the potential benefits of regular PA is evidence of this. The three key themes identified from this research provide insight that has not been previously captured in terms of how the nuances of HD impact on PA experiences and participation across the stages, the importance of the social environment of the person with HD to participation and how people adapt and adjust their PA over time. Furthermore, application of a theoretical model, namely the self-regulation model (SRM) has been used for the first time to facilitate understanding of PA in the context of HD.

It is apparent from the FG data analysis that the SRM is highly relevant and appropriate to help further understanding of PA in the context of HD. The SRM is relevant in terms of the constructs comprising it and the ongoing cyclical nature of regulation that it describes. Development of cognitive representations, responses to perceived health threats and appraisal of response as a continuous cycle described by the SRM are relevant to the progressive nature of HD; the findings suggest that representations of HD and coping reactions with regards to PA continuously alter in response to the changing disease. Self-regulation of PA becomes more of a collaborative regulation between caregivers and people with HD, as with progression, caregiver support becomes increasingly vital to the everyday activities of the individual with HD.

Examples of physical support to exercise, prompts and reminders and strategies to adjust activities are some of the ways that caregiver input was described. In consideration of this, a modified version of the SRM to facilitate understanding of PA in the context of HD has been suggested, and is an original contribution to knowledge as a result of this research.

The proposed adapted self-regulation model specific to PA in HD (figure 17) includes 'moving forward in time' as an element for consideration, coupled with the input from the caregiver and regulation of PA becoming more collaborative between the caregiver and person with HD. The original SRM enables consideration to be given to time because of the cyclic nature of the model. However, specific to HD and PA, the notion of progression of time is linked to an increase in caregiver input and collaborative regulation. This is because the disease becomes progressively and globally debilitating over time. There is a causal relationship between time and caregiver input / collaborative regulation of PA. This may create problems for people if caregivers are not engaged or supportive. Further research that focusses on the role of the changing relationships with progression over time in the context of PA participation could further elicit the impact of changing roles within HD families on PA for the person with HD.

Clinical implications and key considerations for physiotherapists and other healthcare professionals (discussed in more detail in 6.3) include the highlighted need for support with PA with HD progression, which has implications for health services. There are a number of reasons for this, including the potential need for healthcare services due to secondary complications of inactivity in HD and ‘revolving door’ patients due to lack of appropriate support. If people do not have the support at home because of the burden already placed on caregivers, options for ‘exercise buddies’ as described in the MS study (Toomey & Coote 2017) may be of value but again has cost and resource implications for health and social care services. The World Health Organisation (WHO) discusses the issue of “restoring or creating a life of acceptable quality for people who suffer from neurological disorders” (WHO 2006) within the context of public health and yet acknowledge the challenges associated with this. The WHO describes challenges as “a lack of policies, programmes and resources for their treatment and management” (WHO 2006). As demonstrated in one of the FGs, social care services can have a very positive impact in terms of facilitating appropriate PA support. Unfortunately, the lack of consistency in awareness of HD and service provision highlighted in the FGs across the UK suggests more could be done to promote and engage HD families in active lifestyles, highlighting a public health issue.

## **6.2 Limitations**

Research participants were people with HD and or family members and or caregivers of people with HD interested in discussing exercise and exercise experiences in a group format and registered with the Huntington’s Disease Association (HDA). HD is identified as a rare disease (Rath & Kelly 2016), with historical stigma attached to diagnosis (Rawlins et al. 2016) and is considered a hard to reach population. It is possible that others who could have contributed a valuable perspective, did not engage with the research. This could include for example, families who are more isolated from lack of outside support and not being known to the HDA. Other studies have found success in recruiting people within hard to reach populations by using a variety of sampling techniques, such as snowball sampling and network sampling (Thompson & Phillips 2007). Although sampling techniques that depend on people with HD utilising a personal network to help with recruitment may prove challenging, the HDA was helpful in the sense of having a network of care advisors who knew numerous families. Perhaps other such charities or support groups could be helpful in such recruitment approaches. This is an issue that could be explored in future research.

Another limitation is that people who may have been interested in participating but not comfortable with joining a group discussion may not have engaged with the research. It is important to realise that that as an inherited condition, stigma of HD may cause

potential participants to be wary of revealing themselves as gene positive if it could have a negative impact on themselves or family members (Williams et al. 2010). It is also important to acknowledge potential selection bias because those who participated in this study had an interest in physical activity.

The FGs consisted of a mixture of people with HD and caregivers. Separate groups of caregivers and people with HD could have resulted in different discussions because it would have eliminated the dynamic between familial caregivers and people with HD. The researcher has observed from their general experiences of working with people with HD that caregivers sometimes limit what they say about noticing symptom progression in front of the person with HD. However, for some participants it was essential that the caregiver was there to advocate and respond for them. An example is of a woman in FG2 who had limited speech but her caregiver was able to expand on and articulate details (whilst checking with the woman with HD). It was also appropriate for caregivers to be present for those at later stages to prompt or help them in getting their point across. A similar argument was put forward by Carlozzi and Tulsky (2013) who conducted FGs with people across the disease spectrum and their caregivers. They included caregivers because of potential cognitive impairments which would be recognised by caregivers who could prompt and help the individual with HD to remember or identify issues (Carlozzi and Tulsky 2013). Clare and Harman (2006) also suggest that inclusion of the caregivers or family members is of value since they might be able to help explain how the person with HD reacts in different situations and provide details.

Regarding data collection, although the researcher attempted to facilitate a balanced group discussion, it is possible that some participants were more vocal than others. This could have been for numerous reasons, from impaired speech to lack of confidence or extra time needed to process the questions and respond. There may have been more stories or perspectives that could have added to the overall picture obtained from the groups. This could have happened in the large pilot FG (FG1) where it was not possible to split the group into two (due to only one small room being available because of unexpected maintenance work). Managing effective group facilitation was a challenge and there were times where people spoke over each other. Although this was a good learning opportunity for future FG preparation, small sections of the audio recording were difficult to hear and so some information was lost despite having the field notes. Going forward, this issue was addressed as much as possible in facilitation of the FGs. At the beginning of each FG the rules were explained (and if needed, were reiterated during the FGs), including that everybody had equal right to

have their story heard and only one person should speak at a time. Facilitators were aware of potential communication issues, so they knew that enough time should be given to people who took longer to respond. Caregivers were also helpful in prompting their partners. Generally, participants within the groups recognised those who struggled with communicating and were courteous towards them in encouraging them to take part in the discussions. Also, the rest of the groups were a more manageable size for FG discussions (table 10).

As with any qualitative research, the balance of power and relationship between participants and the researcher may be a limitation if participants feel limited in what they can disclose (Karnieli-Miller et al. 2009). In line with the ethical considerations, the researcher addressed this by clearly explaining the research fully to the participants before the FGs, answered any participant questions and explained how the findings would be used. In addition, before beginning each FG there was time for an informal 'meet and greet' with refreshments to help put participants at ease. The use of recording devices can make participants self-conscious (Morse & Field 1995), which may also affect what they say because of self-censorship. The researcher ensured that all participants knew the discussions were being tape recorded, but tried to ensure that the Dictaphone was unobtrusive and not in participants' direct line of sight as much as possible.

The researcher acknowledges that construction of discussion and shared experiences by the FG participants are their interpretations of prior experiences which could be subject to different interpretations over time. Also, how participants chose to depict experiences could have been influenced by social factors including opinions of other FG participants and the picture that participants would like to portray of themselves to others and the facilitator(s) who were physiotherapists. It is also important to acknowledge that, from inception to completion, the researcher's background and perspectives could have influenced the research. The researcher consciously focussed on what was in the data and not making assumptions based on their knowledge or personal experiences (Spinelli 2005). In addition, the theoretically driven analysis may have helped to bracket the influence of the researcher's perspectives on the research findings, avoiding interpretation of the findings in a certain way.

Although the research has captured the experiences of people with HD from prodromal to late stage HD, the method used limited the capture of experiences of those at end stage. It would not be appropriate to invite people at end stage HD to take part in FGs; the severity of impairment by end stage means that full nursing care is required

movement beyond the confines of a nursing bed is unlikely. Speech and meaningful functional movement and cognition are almost completely impaired. An assumption that almost no PA is engaged in at this stage may be challenged by using qualitative exploration and observing interactions between caregivers and those with end stage HD. At this stage, reducing pain and maximising movement through passive movements, active, active assisted movements, positioning and passive joint range of motion may be considered a type of activity. Future research in this area could complete the picture of the full life cycle of HD in an activity context.

### **6.3 Implications for research and clinical implications**

An informed programme of research development, along with the data and knowledge generated from this research will be critical to support development of physical activity (PA) based interventions in HD. These may be in the form of PA programmes, exercise interventions in research, training materials for staff delivering such interventions and supportive resources.

It is important that existing knowledge gained from the interventional studies of exercise in HD that have shown various approaches including inpatient rehabilitation, supported exercise in the community and at home are acceptable and feasible for people with HD, (Zinzi et al. 2007; Khalil et al. 2012; Kloos et al. 2013; Busse et al. 2013; Quinn et al. 2014; Frich et al. 2014; Quinn et al. 2016) are combined with the findings from this research. Increased collaborative regulation could be seen in terms of whoever is involved in supporting the PA of the individual providing verbal feedback as to how activities are performed or encouraging use of different strategies to participate successfully. From a physiotherapy perspective, the evidence from this and previous research indicates that input is needed to encourage physical and functional activity, but a moderated approach would be helpful i.e. knowing when to step in and be more “hands on” in terms of treatment approaches and when to stand back. Quinn et al. (2016b) carefully considered facilitation of translation from research into practice when developing the ENGAGE-HD intervention. The successful use of self-determination theory (SDT) to underpin a PA intervention in HD (Quinn et al. 2016b), suggests that communication styles promoting PA may be complementary to interventions and could also be underpinned by the modified SRM. Modification of the SRM specifically in the context of HD and PA is helpful to reflect the life span of the disease and the changing roles of the person with HD and their caregiver (figure 17). This is an important topic for future research as the importance of PA has been

recognised and understanding how HD changes over time could help develop interventions that evolve and maintain relevance over the life cycle of the disease.

As discussed in the literature review, the MRC framework for development and evaluation of complex interventions (2008) emphasises the importance of qualitative research to achieve better developed interventions and better designed evaluations. The contextual factors, experiences and perceptions of those with HD identified through this research could inform or be further explored through future interventions that incorporate the findings as design considerations (Moore et al. 2015). As the SRM describes, experiential and verbal information received can become integrated into the individual's cognitive representations of their illness. Future research might focus on exploring the merits of targeting cognitive representations to facilitate positive perceptions of PA that encourage participation in the context of HD. Indeed, the findings from this research can build upon the previously discussed ENGAGE-HD intervention (p57-60) in terms of the implementation mapping process which facilitates development of complex interventions based on a foundation of theoretical, empirical and practical information. In discussing the ENGAGE-HD intervention within the literature review, the researcher highlighted that in line with 'implementation mapping' described by Bartholomew et al. (2016), identifying determinants of behaviour was something which may have enhanced the ENGAGE-HD intervention. From the findings of this research, the researcher was able to map their findings to the ENGAGE-HD intervention as an example to demonstrate how the findings from this research build on the existing ENGAGE-HD logic model.

The findings reported here could underpin and enhance development of a wide range of PA interventions in HD in the future. This is because the findings relate to experiences of PA in HD in general i.e. provide insight into the wider context of PA and are not just thoughts specific to an intervention, and were captured across the stages of HD. The first stage in implementation mapping of complex interventions is to develop an understanding of the population that the intervention targets and from this, behaviour determinants can be identified. In figure 18, the researcher illustrates how the research findings relate to components of the 6 step process of implementation mapping to be informative to intervention development by identifying determinants of behaviour and how these relate to SRM. The model clearly depicts what the determinants are (first column), how they were identified (second column) and the components of the identified theory that the determinants relate to (third column). Clearly mapping the components of a complex intervention where there are various interacting factors allows identification of those that work well or do not during the

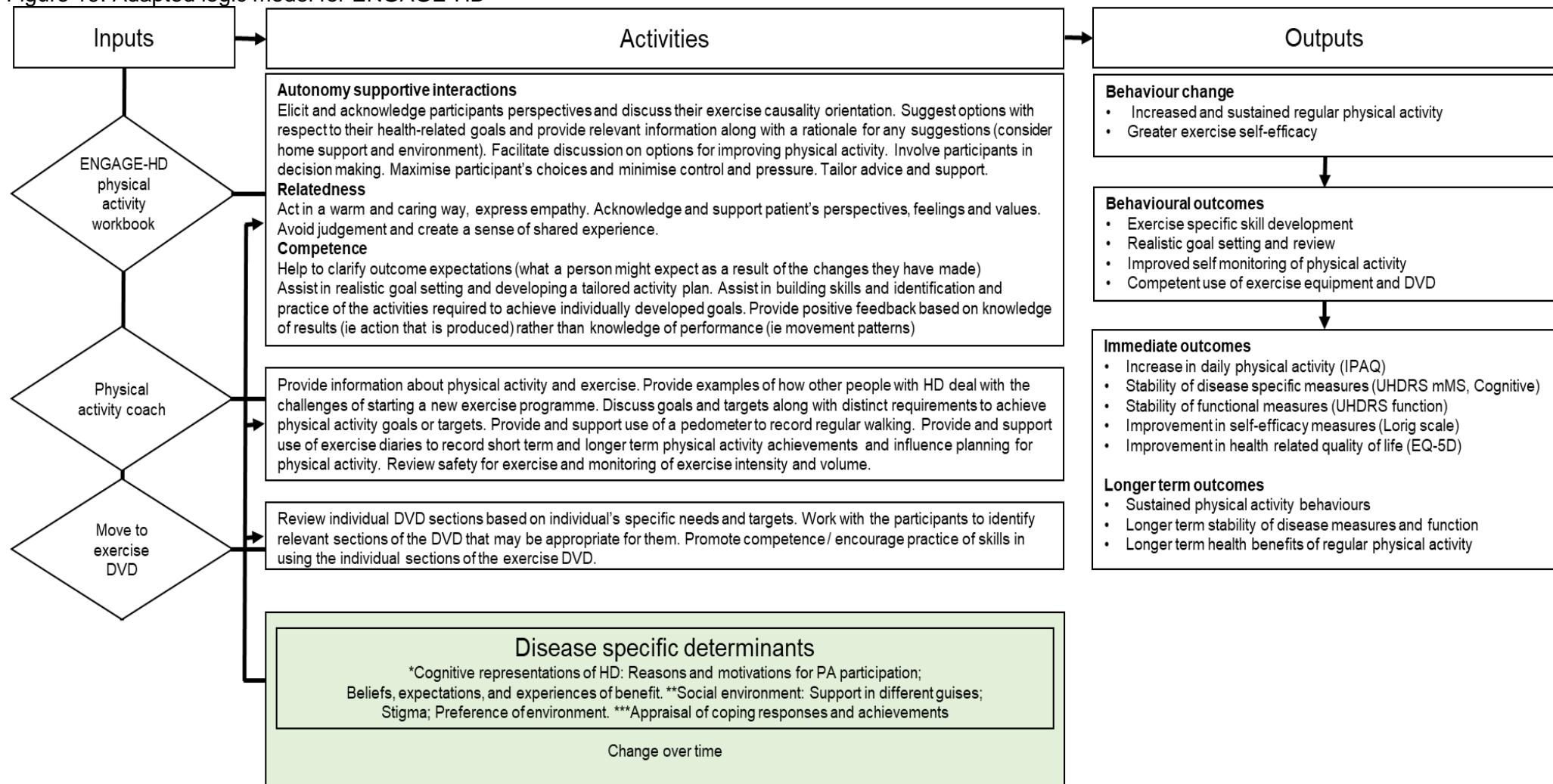
research, which can then be addressed to aid in successful implementation in translating to clinical practice.

In figure 19, an adapted logic model is presented showing determinants of behaviour and behaviour change techniques for ENGAGE-HD as an example of how the findings from this research could build on the theoretical foundations for the ENGAGE-HD intervention. The original contribution from this research is shaded green, the rest is taken from the ENGAGE-HD logic model. The researcher has mapped the themes of this research to the ENGAGE-HD intervention to show how it can build on the theoretical underpinnings of the intervention. The content within the column 'disease specific determinants', which relate to the key themes of this research are all influenced by change over time (indicated in the sidelong box which extends across all of the other determinants). In the approach, the researcher suggests, the FG findings are integrated throughout the whole process ensuring integration of theoretical underpinning throughout development of the intervention.

Figure 18: Determinants of PA behaviour and ENGAGE-HD intervention components

Determinants of PA behaviour	Needs assessment component that provided evidence of the determinant	Self-regulation model (SRM) component	Intervention components and interactions designed to promote motivation for PA underpinned by SDT theory
Cognitive representations of HD:  - Reasons and motivations for PA participation  - Beliefs, expectations, and experiences of benefit	Focus groups around the UK with HD families and caregivers	Perceptions, illness representations	<b>Home visits where coaches interact with participants to promote:</b> <i>Autonomy</i> <ul style="list-style-type: none"><li>- Involving participants in decision making</li><li>- Maximising choices and minimising control and pressure</li><li>- Tailoring advice and support to the individual</li></ul> <i>Relatedness</i> <ul style="list-style-type: none"><li>- Acting in a warm, caring way</li><li>- Acknowledging and supporting patient perspectives, feelings and values</li><li>- Avoiding judgement or blame</li></ul> <i>Competence</i> <ul style="list-style-type: none"><li>- Helping to clarify outcome expectations</li><li>- Assisting in realistic goal setting</li><li>- Assisting in building skills and aiding with activities required to achieve goals</li><li>- Providing positive feedback</li></ul>
Social environment:  - Support in different guises - Stigma - Preference of environment		Coping, illness representations	<b>A purpose-designed workbook for HD patients, which aims to:</b> <ul style="list-style-type: none"><li>- Provide information about the benefits of physical activity and exercise</li><li>- Provide examples of how other HD patients deal with challenges of PA</li><li>- Introduce patients to goal setting, exercise diaries and pedometers for self-monitoring</li><li>- Encourage safety in exercising</li></ul>
Appraisal of coping responses and achievements		Integration of perceptions with memory, Awareness, Coping, Evaluation	<b>A DVD for patients which aims to:</b> <ul style="list-style-type: none"><li>- Give participants a step-by-step guide to performing a range of different exercises in the home, depending on interests and needs</li></ul> <b>Additional complementary ways of supporting participant engagement in ENGAGE-HD intervention</b> Specific individualised strategies for remembering to do PA or adapting to abilities, adapting activities and expectations over time, healthcare professionals being inclusive of caregivers, influencing cognitive representations of PA and HD through education about exercise, and being aware of changes over time, adapting approaches as necessary.

Figure 19: Adapted logic model for ENGAGE-HD



\*relates to KEY THEME 1: The evolving representations of HD and physical activity 207

\*\*relates to KEY THEME 2: The varying social environment of the person with HD and the impact on physical activity

\*\*\* relates to KEY THEME 3: Achieving physical activity participation while coping with the nuances of HD

The framework method facilitated a clear process for the analysis of the FGs and allowed for appropriate exploration of the data. By developing the analytical coding framework using both open coding and the Self-regulation model (described in the methodology chapter), analysis was not narrowed by having a theoretically driven focus only. This method allowed the researcher to explore the emerging patterns within and across the FGs, and develop the key themes. The framework method of analysis would be appropriate for future research in the context of exploring PA experiences in HD. As this research has suggested a modified SRM within the context of HD and PA, further testing of this model to explore its utility and application to help understand more about the experience of living with HD will be important. Given the similarities between HD and some other neurodegenerative diseases, it may be informative to consider exploration of experiences of conditions such as PD or MS using the modified SRM as a lens through which to do so. Such investigation may help to provide insight as to whether self-management or more collaborative management of PA is more realistic for people with PD or MS and identify improvements for structure to clinical care in terms of when people need more help and support.

The overarching concept of change over time was identified as a salient feature of HD that had great impact on PA. 'Change over time' refers to symptom progression over the life course of the disease and the changing identity of the person with HD, both of which have consequences for changing roles within families. This was clearly described by FG participants. Overall, due to HD progression, changes in expectations and priorities, motivations and reasons for exercising altered over time. Consequences of symptom progression include changes in abilities and behaviour; barriers to PA such as apathy, fear of falling, unstable gait and poor balance are seen (Kirkwood et al. 2001; Rosenblatt & Leroi 2000; Wheelock et al. 2003; Goldberg et al. 2010; Collett et al. 2014). Other things which may change over time include confidence to do PA, what people feel comfortable doing, and where, because of the stigma of the visible movement disorder. These changes were described in terms of how people's participation was affected and how people with HD and their caregivers dealt with that across the stages. For example, at earlier stages people found ways of overcoming barriers through using strategies to continue the same activities. However, once gait, balance and cognitive impairments affected their activity, people would adapt activities or do more functional type activities.

Additionally, people removed themselves from exercising in environments where they felt socially awkward because of others staring at them. This has relevance to the self-regulation model (SRM) and physiotherapy input where ongoing adaptation and

appraisal of physiotherapy approaches over time should be incorporated in line with the specific abilities and needs of the individual in terms of advice or suggestions for PA. Redefining one's self in response to limitations and impairments has been identified in MS (Eriksson et al. 2013) and relates to the concept of adjusting and adapting over time. For people with HD, an awareness of how things may change over time from early on and hearing about how others in a similar situation have coped could be a valuable source of informational support. Later on, limited insight may be a challenge to people with HD acknowledging changes, which is where support of not only caregivers and family members but also healthcare professional input can be very valuable.

The FGs highlighted the importance of healthcare professionals, exercise trainers and caregivers being aware of HD symptoms and issues and how changes might affect PA. Across the FGs participants described experiences where healthcare professionals lacked understanding and awareness of HD. Physiotherapists and other clinicians who support people with HD to be active need awareness of the potential changes over time. This is important when designing clinical interventions or giving advice/education, to ensure treatment remains relevant over time and disease course. Drawing attention to the physical and communication difficulties that people with HD may have via information sheets aimed at healthcare professionals could be very useful when developing a plan with patients. In addition, information including potential barriers to PA and strategies could help prepare staff to anticipate and problem solve such issues. The researcher has highlighted the need to accommodate changes in a 'Physio Works' document; an evidence based document for the Chartered Society of Physiotherapy that the researcher was heavily involved in developing (see appendix 22). The document is based online and is used as a reference for physiotherapists who have contact with people with HD for suggestions in their practice. The information contained in the document is targeted towards physiotherapists but there is advice within the document that can be passed onto patients or used to help explain the physiotherapist's suggestions for PA.

Physiotherapists could help to encourage participation collaboratively developing sustainable PA routines. This may be achieved by adjusting the activities they recommend, the environments in which people with HD are active and the advice that they give. As well as helping develop a plan for the actual exercises /activities, approaches could include educating about strategies to motivate or reminders to do the activities. Indeed, Quinn et al. (2010) and Frich et al. (2014) found that participant motivation was positively influenced by understanding the purpose of exercises, goal setting and surpassing previous achievements, and participant collaboration in

developing a personal exercise programme rather than being given a prescribed routine.

Insight was gained from this research into the evolving role of the caregiver and how they support the person with HD as well as strategies that caregivers have developed for lack of support from elsewhere. Certainly, following some of the FGs, caregivers commented that the FGs had been enlightening in terms of sharing and learning about different PA experiences in HD, particularly strategies for engagement. The FG participants reported frequently how helpful it was to hear about others in a similar position, and how they dealt with the impact of HD on their PA participation. Indeed, the social benefits of PA in terms of interacting with others with HD feeling a better sense of control in managing their HD through PA has been highlighted by participants in interventional HD studies (Zinzi et al. 2009; Frich et al. 2014). Modelling positive behaviour of others may also occur through exposure to seeing how others with similar difficulties and abilities succeed in PA engagement as has been described in MS (Rothing et al. 2014). The SRM suggests that integration of information from various sources is one of the contributing factors in the ongoing development of illness representations. If HD families (i.e. the individual with HD and their caregiver/s) receive positive information regarding PA, this may help develop positive representations of how they can use PA to positively influence HD and their well-being. The information could be in the form of information leaflets, advice directly from the physiotherapist or signposting to online resources that demonstrate safe, appropriate exercises relevant for the individual following the physiotherapist's assessment of them. Using resources such as support groups or online resources may be important for building positive representations of PA in HD through direct social communication or the wider surrounding culture through appropriate online resources. Informational support was identified as a facilitator of exercise in PD, MS, motor neurone disease and muscular dystrophy (Ravenek et al. 2009; Elsworth et al. 2009). Ravenek et al. (2009) described the advice from a participant's neurologist to participate in a particular exercise regime as informational support which may have influenced their decision to participate in PA.

An example where information support is utilised is with the Expert Patient Programmes (EPP) Cymru and Expert Patients Programme Community Interest Company (EPPCIC). These programmes use techniques to empower patients to self-manage with part of the 'training' dedicated to educating patients so that they better understand the condition that they have. Profound examples were given by FG participants with early stage HD who used PA as a way of empowering themselves to feel in control and 'fight back' against the disease. The participants believed that they

were doing something positive for their health. Furthermore, participants of this research and previous research expressed what an ‘isolating disease’ HD is and that participation was facilitated by understanding of the potential benefits of PA (Quinn et al. 2010; Frich et al. 2014).

The social environment can also impact on PA for people with HD which speaks to the SRM in terms of how it suggests the surrounding culture and direct social contact inform illness representation development and therefore guide action or coping responses. The social environment of the gym was an important factor to participants of a community exercise programme in HD (Debono et al. 2012) and was noted as a problem for people with MS (Plow et al. 2009). The barriers experienced in terms of the visible disease and social stigma people have faced were also elicited by FG participants. Consideration of such social barriers are crucial for healthcare professionals when designing a treatment intervention or encouraging and problem solving with patients to overcome those barriers. This links to the ideas of the behaviour change wheel (BCW) which suggests that for interventions for behaviour change to be successful they should consider capability (physical or psychological ability), motivation (reflective and automatic mechanisms that activate or inhibit behaviour), and opportunity (physical and social environment) (Michie et al. 2011). The FG data suggests that some barriers may be overcome using simple strategies to continue with the same activities, but in later stages, adaptation of physical activities may be more appropriate to consider. Simple but effective ideas emerged from the FGs that had potential to maintain participation, such as caregivers ‘planting the seed of PA a few days before’ or using a calendar on the wall to write prompts as reminders for people with HD. Goal setting was identified as being helpful for participation, in line with findings from previous studies of PA in HD (Quinn et al. 2010; Frich et al. 2014). Such strategies may become integrated into coping responses to help continue PA in the context of HD.

Another clinical implication relates to considering the social landscape of the individual with HD. Such an approach may be to include caregivers in setting goals, listen for strategies that the caregivers have used with success, or suggest specific strategies that could be used for motivation / reminders. Certainly the success of the ENGAGE-HD PA intervention suggests that tailoring of approaches to the individual to support PA is vital to participation (Busse et al. 2017). Through the knowledge and understanding gained from this research, the researcher has been able to consult on further development of the ENGAGE-HD workbook (version 2) that builds on the success of the original (Busse et al. 2017). Self-management approaches that include

PA are vital as PA has been indicated as a potential disease modifying intervention. Self-regulation plays a key part in this and further consideration of the findings presented is that the regulation of activity for the person with HD becomes more collaborative with the caregiver over time. As Rothing and colleagues report (Rothing et al. 2014), it is important for caregivers to be involved when creating exercise programmes or routines, as they become more integral to the patient's PA participation with progression. If caregivers are not on board, education and support for them may be important.

Early intervention is another important consideration for a progressive, long term disease which, as yet, has no cure. The findings presented here suggest that regular PA participation from an early stage may be beneficial in terms of the social opportunities for people with HD, developing long term habits of PA and maintaining functional abilities for as long as possible in line with findings of interventional studies (Zinzi et al. 2007; Frich et al. 2014). Engaging in PA from an early stage and understanding the potential benefits may be important for well-being, may empower patients to feel in control and maintain function and independence for longer through keeping active. If people with HD were to develop PA behaviour as part of their lifestyle from early on, prior to the issues associated with progression of the HD such as the movement disorder, social stigma or embarrassment and apathy, they may continue and adapt as needed as the HD progresses. Explaining to patients and family members that they will need to adapt and respond to changes due to HD may help their understanding and awareness that the individual with HD may need increased support to appraise and adapt their activities.

Disseminating the findings in such a way as to highlight the positive ways that people with HD are able to engage in PA could therefore be helpful for HD families. As such, the researcher to date has used the findings in development of an online information resource to support PA for HD families and formal caregivers. This was possible through her work as co-chair of the European Huntington's Disease Network Physiotherapy Working Group. Understanding gained from the findings of this research has also helped the researcher conceptually develop a project aimed at developing walking groups for people with HD.

## **6.4 Final conclusions**

The aim of this research was to explore how living with HD impacts on the experience of PA across the stages of the disease. HD is a unique disease encompassing physical, behavioural, cognitive and psychiatric symptoms that progress over time and is presently incurable. Everyday living with HD should include planning for PA because of the potential physical, social and psychological benefits gained from regular engagement. This is particularly poignant in a disease where the needs of people with HD are not always met because of lack of healthcare professional or caregiver understanding of the disease or effective medication to slow disease progression or delay onset (Novak & Tabrizi 2010; Skirton et al. 2010). The growing evidence base that favours being physically active in HD means it is important that healthcare professionals and caregivers can support people with HD to engage in PA (Quinn et al. 2013). It is therefore important to understand personal experiences of PA in HD to explore how the nuances of the disease impact on participation.

The experiences and perspectives that research participants shared has furthered understanding, that for people with HD, change over time is an important feature that impacts different aspects related to PA. The self-regulation model (SRM) was integral in facilitating understanding of the impact of HD on PA in terms of the relevance of the process of regulation that people go through and the constructs of the illness representations including identity, coping, control and consequences. In comparison to the ongoing cycle of self-regulation described by the SRM, a slightly modified concept of ‘collaborative regulation’ of representations and PA participation was seen with progression of HD. The findings concur with that from previously published studies that describe changing roles within the family unit and the continually evolving nature of the HD. In line with findings from process evaluation of specific exercise interventions in HD, similar facilitators and barriers were elicited such as social stigma and individual support. The research findings additionally give insight into how PA participation is achieved in the face of coping with the nuances of HD through to late stage, giving insight into the strategies used and adjustments made by people with HD and their caregivers. Due to the method used, the research does not capture the experiences of those at end stage who are bed-bound and completely dependent on full care assistance. This is a consideration for future research that could look at exploring activity related to function at end stage.

Potential directions for future research have been suggested, and implications for clinical practice. Given the potential benefits that may be acquired through regular PA, the general feeling of participants that there is lack of awareness and provision for

appropriate PA support in health and social care generally, indicates a public health need and is something that should be addressed urgently. Primarily, the clinical implications relate to consideration of changes over time in HD when designing clinical interventions or giving advice/education, the social landscape of the person with HD, early intervention to encourage PA participation from an early stage, and at later stages the benefit of doing little and often and perhaps taking smaller steps to achieve realistic goals.

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## Appendices

## Appendix 1: Summaries of studies exploring treatment approaches in Huntington's disease

Title	Author / date	Study design	Aim	Sample	Type of intervention for experimental group	Type of intervention for control group	Outcomes	Results
A randomized, controlled trial of a multi-modal exercise intervention in Huntington's disease	Quinn et al. 2016	Randomised controlled feasibility study	To evaluate the feasibility and benefit of a structured exercise intervention in people with Huntington's Disease (HD).	Thirty two individuals with Huntington's disease who were able to use a stationary bike, had a stable medication regime for four weeks prior to initiation of trial, and anticipated to be able to maintain a stable regime for the course of trial and did not have any physical or psychiatric condition that would prohibit the participant from completing the intervention or the full battery of assessments, were not currently involved in any intervention trial or within four weeks of completing an intervention trial or currently participated in a structured exercise program five times per week or more.	The intervention was 12 weeks, three times per week progressive exercise program, including aerobic (stationary cycling) and upper and lower body strengthening exercise with tapered 1:1 support for 20 of 36 sessions.	Continue as normal, diaries given – no other input.	The primary efficacy outcome in terms of short-term benefit was physical fitness measured using a predicted VO <sub>2</sub> max equation[13], by stepwise incremental exercise test. The primary feasibility objective was evaluation of recruitment, retention and adherence rates.	The intervention group had statistically significantly better fitness (predicted VO <sub>2</sub> max difference: 493.3 ml.min <sup>-1</sup> , 95% CI: [97.1, 887.6]), lower UHDRS mMS (difference 2.9 points, 95% [-5.42, -0.32]) and lower weight at Week 13 (difference 2.25 kg, 95% CI: [-4.47, 0.03]). Three individuals (intervention group) were withdrawn within the first month due to concomitant medical conditions (Figure 1), resulting in 14 participants in the intervention and 15 in the control group for final analysis (retention rate of 90.6%, 95% CI [73.4, 97.5]. All assessors remained blinded throughout the study. Thirteen of the 14 participants who completed the trial completed >75% of the required sessions (92.9%, 95% CI [64.2, 99.6]); one participant completed 61% of sessions secondary to illness. For the aerobic exercise, 10/13 achieved average target HR within aerobic zone (65-85% APMHR) for at least 75% of the sessions.
Task specific training in Huntington's disease : a randomized controlled feasibility trial.	Quinn et al. 2013	Randomised controlled feasibility study	To assess feasibility and safety of goal-directed, task-specific mobility training for individuals with mid-stage HD.	Thirty individuals with mid-stage HD (13 men, 17 women; mean age=57.0 years, SD=10.1)	Task-specific training was conducted by physiotherapists in participants' homes. Training focussed on walking, sit-to-stand transfers, standing, twice a week for 8 weeks. In order to individualise the intervention and monitor achievement of personal goals goal attainment scaling was used.	Continue as normal – no other input.	Adherence and adverse events were recorded.	Loss to follow-up was minimal (n=2); adherence in the intervention group was excellent (96.9%). Ninety-two percent of goals were achieved at the end of the intervention; 46% of the participants achieved much better than expected outcomes. Effect sizes on all measures were small.
A randomized feasibility study of a 12-week community-based exercise program for people with Huntington's disease.	Busse et al. 2013	Randomised controlled feasibility study	Assessed feasibility, safety, acceptability, and benefit of a structured exercise programme.	People with early-mid stage HD n=31 (males n=16)	A weekly supervised gym session of stationary cycling and resistance exercises, and a twice weekly independent home-based walking program for 12 weeks.	Continue as normal – no other input.	Measures of strength, mobility, balance, gait, cognition, quality of life, function, disease specific measures (UHDRS functional assessment, problem behaviours assessment, total motor score).	Significant difference in a mental health component of a quality of life questionnaire (SF-36).
Video game play (Dance Dance Revolution) as a potential exercise therapy in Huntington's disease: a controlled clinical trial.	Kloos et al. 2013	Cross-over controlled single blind study.	To investigate the feasibility, acceptability, and safety of a supervised video game exercise program administered via Dance Dance Revolution in individuals with Huntington's disease.	People with HD (stage not specified) n=20	Dance Dance Revolution game with supervision and the handheld game without supervision for 45 minutes, two days per week for six weeks.	Cross-over design – during control condition participants continued as normal without any other input.	Balance: Tinetti Mobility Test-balance section, Four Square Step test.	Significant improvement was observed for double support percentage forward (2.54% reduction, p = 0.03) and backward walking (4.18% reduction, p = 0.01). In evaluating intervention effectiveness depending on severity of motor impairment, a significant interaction was observed for forward heel-to-heel base of support (p = 0.05).

Title	Author / date	Study design	Aim	Sample	Type of intervention for experimental group	Type of intervention for control group	Outcomes	Results
Effects of an intensive rehabilitation programme on patients with Huntington's disease: a pilot study.	Zinzi et al. 2007	Individual case control study	To investigate the effects of an intensive, inpatient rehabilitation programme on individuals affected by Huntington's disease.	People with early to mid-stage HD (n=40)	respiratory exercises and speech therapy, physical and occupational therapy and cognitive rehabilitation exercises. The programme involved three-week admission periods of intensive treatment that could be repeated three times a year.	No control group	Mobility and functional status; Tinetti Mobility Test-gait score, Physical Performance Test .	Each period of treatment resulted in highly significant ( $P<0.001$ ) improvements of motor performance. The average increase was 4.7 for Tinetti and 5.21 for PPT scores. There was no apparent carry-over effect from one admission to the next; however, no motor decline was detected over two years, indicating that maintenance of a constant level of functional motor performance in patients was achieved.
Effects of a One Year Intensive Multidisciplinary Rehabilitation Program for Patients with Huntington's Disease: a Prospective Intervention Study	Piira et al. 2013	Prospective intervention study	To assess the effects of an intensive, multidisciplinary rehabilitation program for patients with early to mid-stage Huntington's disease.	People with early to mid-stage HD (n=37)	A one year rehabilitation program, consisting of three admissions of three weeks each, focus on physical exercise, social activities, and group/teaching sessions.	No control group	Standard measures for motor function, gait and balance, cognitive function, including Mini Mental State Examination and UHDRS cognitive assessment, anxiety and depression, activities of daily living (ADL), health related quality of life and Body Mass Index (BMI).	Significant improvements observed in gait function, balance, in physical quality of life, anxiety and depression, BMI. ADLfunction remained stable with no significant decline. Cognitive measure symbol digit modalities test (SDMT) showed significant decline, no decline was observed for other cognitive measures.
Physical therapy for people with Huntington disease: current perspectives and case report.	Quinn et al. 2002	Single case study	Assessed effects of a home based exercise programme in the form of an exercise video.	Person with mid-stage HD (n=1)	Home-based programme exercise video; balance and muscle strength.	No control group	Disease specific motor score; UHDRS-motor Balance; the Berg Balance Scale. Mobility; gait speed, falls efficacy scale. Quality of life; SF-36.	Improvements in all outcomes; balance, walking speed, decreased number of falls, improvement in the quality of life scale.
Physical therapy for patients with Huntington's disease: effects of a treatment program and intercorrelation between outcome measures.	Ekwall et al. 2010	Individual case-control study	Evaluated the effect of a physiotherapeutic exercise program for patients with HD concerning motor function and disability, balance and fall related self-efficacy.	People with early to mid-stage HD (n=12)	Physiotherapy aimed to improve motor function, disability, balance, fall related self efficacy, focus on transitions, walking, balance, posture, postural control, fall related self efficacy.	No control group	Disease specific motor score; UHDRS-motor Balance; Berg Balance Scale, one leg stance test, figure of eight test Mobility; Timed Up and Go test, the falls efficacy scale.	Improvement in balance measured by Berg balance scale ( $p=0.045$ ) positive effects on motor function ( $p=0.076$ ), and fall related self efficacy ( $p=0.089$ ).

## Appendix 2: Summaries of studies of experiences of the personal experiences of living with Huntington's disease

Study	Population	Exposure	Outcome	Key significant findings
Arran et al 2014	People with HD (n=87) completed the questionnaire online or by post (female n=49; male n=38)	Huntington's disease	The Illness Perceptions Questionnaire was used to explore the biopsychosocial factors related to psychological distress in people with HD, and examine the relationship between illness perceptions, coping and psychological distress. Self-report measures completed by participants related to coping and psychological distress (SF-36 health related quality of life questionnaire and Hospital Anxiety and Depression Scale (HADS)) and data with regard to clinical and demographic variables were collected that are associated with psychological distress.	Illness representations reflect distinct and meaningful constructs for people with HD. In relation to the illness representation domain of identity, fatigue (reported by 65.5%), sleep disturbances (64.4%) and loss of strength (62.1%) were most commonly reported. Most commonly reported HD specific symptoms were involuntary movements (82.8%), problems with balance (80.5%), problems with concentration (79.3%), memory difficulties (72.4%), problems with walking (71.3%), difficulties reading and writing (69%) and personality changes (60.9%). Participants perceived HD to be chronic, as opposed to cyclical and to have strong, negative consequences. Scores on the control domains (personal and treatment) showed that participants perceived little perceived control over the HD. The SF-36 results indicated perceptions of limitations in performing all physical activities, physical health causing a high number of problems with work or other daily activities greater pain in comparison with the general population. The HADS indicated that approximately 48% of individuals possibly had anxiety (n=42) and 40% possibly had depression.
Aubeeluck et al 2012	Family member caregivers (n=47); 26 spousal; 21 'other' family members	Huntington's disease	Six semi-directed focus groups were used to explore impact of HD on quality of life for family caregivers of people with HD.	Four superordinate themes were identified from analysis: 'Levels of Support', 'Dissatisfaction with Caregiving Role', 'Practical Aspects of Caring' and 'Feelings and Emotional Well-being'. Quality of Life is compromised HD family caregivers. Their own care needs were negated; their caregiving role overwhelmed them and 'took over' their lives. Lack of services, understanding from professionals, secrets within the family, isolation were some of the subthemes identified that contributed to the overall picture.
Carlozzi and Tulsky 2013	Individuals with HD (n=24 across the HD spectrum), individuals either at-risk or prodromal HD (n=16) caregivers (n=17), HD clinicians (n=25)	Huntington's disease	In order to develop domains for a HD specific health related quality of life (HQOL) patient reported outcome measure, Carlozzi and Tulsky (2013) conducted focus groups. Sixteen focus groups were conducted, (n=6 groups with symptomatic HD individuals; n=5 with individuals who at-risk or prodromal HD; n=3 non-clinical HD caregivers; n=2 groups with HD clinicians). Semi-structured focus group guides were developed to assess participants' health related quality of life experiences in order to help develop a HD specific quality of life patient reported outcome measure. The focus groups explored quality of life issues relevant to people with HD. Physical, emotional, social and cognitive health were found to be important domains to people with HD. In particular, the importance of constructs related to emotional and social health was identified across the spectrum of HD. To ensure that the HQOL domains developed from the findings would cover all appropriate areas for clinical intervention, clinicians were also included in separate focus groups (speech therapists, nurses, physicians, psychologists, social workers, physiotherapists, dietician, recreational therapists and nursing assistants).	Analysis was carried out using a codebook to code the data that was developed using the multidimensional HQOL theoretical framework developed by the World Health Organization which includes physical, social, and emotional well-being, PROMIS, Neuro-QOL and the data itself. Emotional health, social participation, physical health, cognitive health, and end of life issues were discussed and authors concluded the importance of developing HD-targeted items to ensure sensitive assessment of health related quality of life in HD research and clinical. HD was reported to impact significantly on social participation (including leisure activities) and social relationships.
Downing et al 2010	7 people with prodromal HD and 6 companions.	Huntington's disease	Semi-structured telephone interviews exploring work function; being on time, relationships at work, quality of work, mood, interest in work, how individuals with pre-HD manage changes in work function and whether they have advice regarding work function for others with pre-symptomatic HD.	Participants made attributions related to health, work, and temperament. Only one participant attributed a change to HD. Symptom monitoring and comparison of participants with pre-HD to others with and without HD was elicited as well as uncertainty regarding how to make attributions. Authors suggest this may have implications for health behaviour as attributions influence coping procedures.

Etchegary et al 2011	Participants included people at risk of HD (n=8), gene positive (n=3), gene negative (n=5), waiting for test results (n=2), manifest HD (n=2), family members, not at risk (n=4).	Huntington's disease	Semi structured interviews were carried out to explore the healthcare experiences of HD families and their suggestions for improvement.	Complex needs in terms of healthcare services and emotional supports were identified with frustration elicited at lack of knowledge from healthcare professionals and difficulty accessing appropriate services. Better education and regular follow up support were suggested for improving the healthcare experience.
Hagberg et al 2011	People gene positive for HD who had received the positive gene test more than 5 years ago (n=10).	Huntington's disease	Semi structured interviews were conducted to explore lived experiences; practical issues (career, education, family planning), emotional and cognitive reactions (acceptance of the test results, thoughts about the disease, feelings of guilt) and social experiences (implications for relationships with other family members, friends and fellow workers)	Experiences of positive and negative impact on the participants were shared, some regretted the decision, others had a greater appreciation of life. Confirmation of the result had either acted as a motivator or barrier to pursuit of further education, career, investment in personal health and tended to bring families closer together.
Hagen 2017	Individuals were: HD diagnosis (n=2), gene positive (n=1), gene negative (n=2), at risk (n=1), family members not at risk (n=5).	Huntington's disease	Semi-structured interviews were carried out with the 11 individuals to explore how they experienced everyday life affected by HD.	Data analysis identified two themes (1) noticing symptoms in everyday life and (2) neither health nor disease. Experiences were found to be dynamic and fluid, the difficulties of knowing one is at risk was highlighted as well as the gradual changes in abilities with progression of HD.
Hartelius 2010	People with HD (n=11), family members (n=7)and carers (n=10)	Huntington's disease	Participants were interviewed in focus groups or individually, using a semi-structured interview guide to explore aspects of how communication is affected by HD.	The key themes identified from data analysis were: Communication has changed; Factors that influence communication negatively; and Factors that influence communication positively. People with HD highlighted a need for a richer social life and more (adjusting) conversation partners, a need for more support was expressed by family members. and professional carers wanted more information about HD.

Helder et al 2002a	Individuals with a clinically confirmed diagnosis of HD (n=77)	Huntington's disease	The Illness Perceptions Questionnaire (Weinman et al. 1996) was used to assess the five components of illness perceptions described by Levanthal et al. (1984): identity, time-line, cause, consequences, and control/cure. This was to help describe illness perceptions and coping mechanisms and their role in the well-being of people with HD.	Many symptoms of HD reported, some negative consequences of the disease for their everyday lives, disease would have a long duration, not much hope for cure or improvement of symptoms, could not state with certainty whether they had control over the disease process and the main cause was considered to be genetic with stress as an additional contributing factor to their illness. No significant differences between male vs female, employed vs unemployed on the consequences scale. Differences were seen in the identity scale between those who had to give up work because of HD and those still working and there was a significant positive correlation between duration of HD and scores on the 'identity' scale and significantly negatively associated with a disease specific motor score. In terms of coping, acceptance was seen more often in HD than in 'healthy control individuals' and sought less instrumental social support, venting of fewer emotions, suppressing fewer competing activities, and adopted fewer strategies to mentally disengage from the stressful situation (HD). Illness perceptions characterised by a strong illness identity were negatively related to patient's well-being.
Helder et al 2002b	Spouses of people with Huntington's disease (n=90)	Huntington's disease	This study investigated how coping mechanisms and illness representations contribute to quality of life in spouses of people with Huntington's disease. Interviews and questionnaires were conducted at the participants' homes. Questionnaires included the Illness Perception Questionnaire, (Weinman 1996), the COPE inventory (assesses coping strategies) and quality of life using the Medical Outcome Study 36 item Short Form Health Survey (MOS SF-36) and the Sickness Impact Profile (SIP) (Bergner, Bobbitt, Carter, & Gilson, 1981).	Spouses of people with HD perceived their partner's HD as a high number of somatic symptoms (strong illness identity), being of long duration, and having negative consequences for their day to day living. They reported having little hope for cure or control of the HD symptoms that they believed were mainly caused by genetic factors. The authors suggest that the coping strategies adopted by spouses were similar to reference participants dealing with everyday stressful situations, therefore HD does not seem to affect their quality of life. However they also point out that the measure used for illness perceptions may have been too global therefore not sensitive to capture specific contextual issues. Devising strategies to deal with HD was positively related to spouses' role functioning. They responded to the HD by accepting the disease and the problems caused by it, took action to solve the problems encountered, and tried to develop strategies to deal with the problems at hand.
Kaptein et al 2007	People with HD (n=36) and their partners (n=19)	Huntington's disease	The Illness perception questionnaire, Unified Huntington's disease Rating scales, mini mental state questionnaire and Medical The Medical Outcome Study 36-item Short Form Health Survey were conducted with patients and their partners, separately at home to investigate whether partners hold similar views about HD, to investigate patterns between illness perceptions of patients and their partners and quality of life and to explore the cognitive factors related to Quality of Life for couples living with HD.	In general, people with HD held more positive beliefs about HD than their partners did and attributed significantly less symptoms to HD compared to their partners. People with HD believed that the disease was more controllable than what their partners perceived. Stronger beliefs in control over the illness and less serious perceived consequences of the people with HD were related to better partner quality of life. For people with HD, a less severe illness identity and longer perceived duration of the illness, less consequences, more control and less belief in treatment were related to a better quality of life. In addition, stronger beliefs of the partners in a longer duration of disease and less belief in cure through treatment were related to higher vitality scores of the people with HD. Perceptions held by the partners of identity and consequences were not significantly related to patients' vitality, social functioning or mental health ratings. Stronger beliefs in control over HD from the people with HD were linked to higher vitality scores of partners. Spousal illness perceptions were related to vitality scores of quality of life for both people with HD and their partners, although their own perceptions were responsible for the largest variance in quality of life.

Maxted et al 2014	7 dyads (person with or at risk or HD and their mother/father/daughter/son)	Huntington's disease	Semi structured interviews were conducted with each dyad to examined the experiences related to relationships with their family and each other.	Key themes were identified as 'A spectre hanging over us'; 'Us against the world'; and 'That could be me in 50 years'. Supportive and protective dynamics between dyads were elicited but also presented challenges for outsiders to access the families; presenting challenges particularly for healthcare providers. The authors suggest the need for increased understanding of the needs of HD families.
Røthing et al 2014	15 family member caregivers (3 male; 12 female)	Huntington's disease	Semi structured interviews were conducted with individuals to explore the impact of HD on roles within the family and structure of the family.	Findings suggest HD has an impact on the stability of family systems where hierarchy within families related to spouses and partners, parents and children and roles within the family change. More knowledge about appropriate interventions is needed from healthcare professionals, particularly in terms of social support as families can become isolated with the social impact and stigma of HD families experience.
Skirton et al 2010	Caregivers of individuals with HD (n=227)	Huntington's disease	The 'Community Health Services Scale' was sent to caregivers of people with HD for completion in order to obtain their perceptions regarding availability and adequacy of health and social care services for their family member.	Concerns were elicited regarding the knowledge of healthcare professionals providing the care and insufficiency of services to support the person with HD and the caregivers themselves. 'Community resources', 'individualized care' and 'knowledge of Huntington's disease' were issues highlighted and it was found that caregivers were likely to have responsibilities for earning and caring for children as well as the person with HD.
Williams et al. 2007	19 family members of people with prodromal HD	Huntington's disease	Focus groups were carried out with adult family members of people who had received a positive gene test for HD, but had not received a clinical HD diagnosis. Perceptions of family members were sought in relation to changes in health and function in their family member at prodromal stage, and circumstances that compromise their attempts to manage such changes.	Family members reported observing alterations in cognition, behaviour, and functioning prior to clinical diagnosis. Concerns about relationships, roles and responsibilities, future caregiving, maintaining spousal relationships, responding to changes and the ability of the individual with prodromal HD to carry out their usual activities. It is suggested that earlier interventions during this period may be beneficial to support families.
Williams et al 2009	42 family caregivers of people with HD	Huntington's disease	Focus groups were conducted in the US and Canada in order to explore the emotional experiences of caregiving by family caregivers of people with HD and describe strategies they used to deal with that experience.	One core theme was identified from thematic analysis 'caregiver's emotional experiences'. Sub themes of this describe experiences of 'disintegration of one's life', how caregivers adapt to loss of life as it was prior to their family member developing HD, and experiences of how they dealt with discussing the gene test with their children who were at risk. Coping was attempted through seeking comfort from selected family members. Spousal carers were distressed by the loss of their spousal relationship. They attempted to deal with this by no longer thinking of that person as an intimate partner. Caregivers attempted to maintain multiple roles, the authors argue that emotional distress can compromise the well-being of family caregivers. Authors suggest these findings indicate mental health of family caregivers should also be monitored, and sources of emotional distress identified and effective strategies used by caregivers to mediate distress should be supported by healthcare professionals.

Appendix 3: CASP summary table: critical appraisal of studies related to experiences of Physical Activity in Huntington's disease

Author, date	Clear statement of aims?	Qualitative methodology appropriate?	Research design appropriate to address the aims?	Recruitment strategy appropriate to the aims?	Appropriate use of theoretical framework to underpin the research?	Was the data collected in a way that addressed the research issue?	Has the relationship between researcher and participants been adequately considered?	Have ethical issues been taken into consideration?	Was the data analysis sufficiently rigorous?	Is there a clear statement of findings?	How valuable is the research ?
Quinn et al. 2010	Yes	Yes	Yes	Yes	No, none used	Yes	Yes	Yes	Yes	Yes	Identified barriers to exercise in early-mid stage HD.
Khalil et al. 2012	Yes	Yes	Yes	Yes	No, none used	Yes	Yes	Yes	No	Yes	Elicited disease specific barriers due to cognitive and physical impairment within the context of a specific intervention.
Zinzi et al. 2009	Yes	Yes	No	Yes	No, none used	No	No	Yes	No	No	Lacked discussion of qualitative findings, limitations associated with postal questionnaire.
Busse et al. 2013	Yes	Yes	Yes	Yes	No, none used	Yes	Yes	Yes	Yes	Yes	Insight into adherence to a community based programme for early to mid-stage HD. Motivation was

											facilitated by perceived benefits.
Frich et al. 2014	Yes	Yes	Yes	Yes	No, none used	Yes	Yes	Yes	Yes	Yes	Provides insight into participant perceptions and experiences of an in-patient rehabilitation programme, particularly aspects that they found helpful such a joint goal setting.
Quinn et al. 2016	Yes	Yes	Yes	Yes	Yes – self-determination theory	Yes	Yes	Yes	Yes	Yes	Exploration of participant perceptions was gained in relation to development of a supportive workbook that was being developed for use in an exercise trial.

Appendix 4: Summaries of studies of experiences of physical activity in Huntington's disease

<b>Study</b>	<b>Population</b>	<b>Exposure</b>	<b>Outcome</b>	<b>Key significant findings</b>
Zinzi et al 2007	People with early to mid-stage HD (n=40) who had participated in the inpatient multidisciplinary rehabilitation programme.	Huntington's disease	Postal questionnaire exploring experiences of an inpatient multidisciplinary rehabilitation programme. The programme consisted of respiratory exercises, speech therapy, physical and occupational therapy and cognitive rehabilitation exercises. The programme involved three-week admission periods of intensive treatment that could be repeated three times a year.	Barriers to completing the rehabilitation programme; lack of motivation, organisational difficulties in attending, ambivalence about staying away from home during the programme. Facilitators: support from family and staff, patient centred approach, physical and psychosocial benefit and social interaction. Participants reported better sense of control in management of their HD, improved self-esteem and better knowledge of HD by participating in the programme.
Quinn et al 2010	People with early-mid stage Huntington's disease (n=5) and Parkinson's disease (n=5). Eight physiotherapists with more than 2 years' experience of working with people with neurological conditions.	Huntington's disease and Parkinson's disease.	Two focus groups with physiotherapists (n=8). Individual interviews with people with Huntington's disease and Parkinson's disease (n=10).	Barriers to engaging in self-directed home exercise included disease-specific limitations, safety and location of exercise. Strategies for motivation were used and included setting targets and having knowledge of benefits of exercise.
Khalil et al 2012	Fifteen participants with HD who had taken part in a home based exercise programme supported by a DVD.	Huntington's disease	Semi-structured individual interviews.	Commitment of the caregiver was a key to the success of the programme. Barriers and facilitators to using the exercise DVD were reported, management strategies helped promote adherence to the exercise programme. Self-efficacy and outcome expectations facilitated participation. (Self-efficacy is a belief in one's capabilities to carry out a given task to attain a desired outcome (Bandura 1998). Outcome expectations are described by Bandura as the expectations that one has about 'the effects of different lifestyle habits', where positive outcome expectations act as incentives for engaging in a health behaviour and negative ones act as disincentives (Bandura 1998)

Frish et al 2014	Nine family caregivers and 11 patients with early- and mid-stage HD, 15 health professionals	Huntington's disease	Interviews with family caregivers of participants and participants of a one year intensive multidisciplinary rehabilitation programme for patients with Huntington's disease. The programme consisted of three inpatient admissions of three weeks each, and a 5 day evaluation stay approximately 3 months after the last rehabilitation admission. The programme focussed on physical exercise, social activities, and group/teaching sessions. Focus groups with healthcare professionals who delivered the programme.	Positive outcomes of participating in the group programme included physical, mental and social benefits. Feeling that the situation was "stable" was important to participants and families. Some participants needed greater support to develop goals as part of their exercise routine and reminders to attend sessions because of cognitive and behavioural difficulties.
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Appendix 5: Summaries of studies of experiences of physical activity (PA) in other neurodegenerative conditions

<b>Study</b>	<b>Population</b>	<b>Exposure</b>	<b>Outcome</b>	<b>Key significant findings</b>
Drugonski et al 2012	Women with Multiple Sclerosis (n=11) who had low levels of disability and engaged in varying levels of PA.	Multiple Sclerosis	Semi-structured interviews (x2) focusing on participant beliefs, motivators, and experiences of PA.	Motivations related to being physically active included the desire to be 'normal', enjoyment, feeling good after activity, weight control, maintaining physical function and savouring current health. Different strategies were used by participants which included prioritising and scheduling PA, managing disease-specific barriers, and building social support networks. Participants described a sense of accomplishment and pride, weight control, health benefits and the importance of a social support network.
Dodd et al 2006	Seven women and two men (mean age 45.6 years) with multiple sclerosis who were participants of a 10-week (2 sessions a week) gymnasium based progressive resistance strengthening programme.	Multiple Sclerosis	Semi-structured interviews were used to explore positive and negative effects of a progressive resistance exercise programme and facilitators and barriers to participation.	Positive factors included physical benefits, social and psychological aspects. Negative perceptions related to some specific exercises of the programme and muscle soreness following sessions. Knowledge of MS of exercise trainers was valued highly.

Elsworth et al 2009	Twenty four people with either muscular dystrophy, multiple sclerosis, motor neurone disease or Parkinson's disease.	Neurological conditions	Focus groups with individuals with neurological conditions were conducted to explore opinions on factors facilitating PA participation. Four condition specific focus groups were carried out. A questionnaire was developed using the themes that emerged from the focus groups and was posted out to people with neurological conditions (n= 80 out of 115 completed) to further cover the topics of necessary support, barriers, and average time spent participating in PA.	Three themes were identified; 'opinions of PA', 'barriers to PA', and 'factors that would encourage increased PA involvement'. Swimming, stretching and walking were the most popular activities. Barriers to participation in PA included embarrassment, perceived lack of condition-specific knowledge of the fitness professionals about neurological disease and the impact of that on exercise advice. Group based exercise sessions (for specific conditions) and the presence of specifically trained staff were perceived facilitators.
Eriksson et al 2013	Eleven individuals (6 males) with Parkinson's disease (PD) who had been regularly participating in exercise for more than 1 year and had participated in a PD specific exercise programme.	Parkinson's disease	Individual interviews to explore experiences and generate understanding of the meaning of exercise for people participating in an exercise programme.	A core category and six categories were identified. The core category related to participants' efforts to maintain function and activities through being active 'keep moving to retain the healthy self'. Other categories related to other concepts considered important to start and maintain PA habits and included categories of "Taking rational position", "Exercising to slow progression", "Exercising to achieve well-being" and "Using exercise as coping strategy". Having goals and self-efficacy were identified as important for adherence.

Smith et al 2009	Ten people (8 females; 2 males) with Multiple Sclerosis who had experienced changes in energy levels since diagnosis but could still walk short distances independently and had participated in a 8-week exercise programme specific to MS.	Multiple Sclerosis	Individual semi-structured interviews were used to explore the influence of an 8-week exercise programme on fatigue perceptions.	Positive and negative perceived influences of exercise on fatigue perceptions were found. Categories identified related to strategies for dealing with fatigue, control over fatigue, outcomes of exercise; physical improvement or deterioration, wellbeing or negative feelings.
Jones et al. 2008	Twenty people with Parkinson's disease (12 male, 8 female; mean age 65 years (range 50 – 80); mean disease duration 10 years (range 2.5 – 26).	Parkinson's disease	Semi-structured interviews were used to explore the personal experience of everyday walking with Parkinson's disease.	Challenges and the strategies used to compensate for difficulties were identified. Findings showed that the physical and psychosocial context of everyday walking need to be accounted for by rehabilitation interventions and care giver input can be important for monitoring and supporting walking activity.
Kayes et al. 2011	Ten people with Multiple Sclerosis (7 females; 3 males); time since diagnosis ranged from 3 -17 years.	Multiple Sclerosis	Semi-structured interviews explored engagement in PA from the participant perspectives to gain a more in-depth understanding of the barriers and facilitators to engagement.	Core themes were identified as "beliefs about PA", "emotional response", "fatigue", "pre-MS activity", and "moderating factors". Strength of beliefs about PA had an influence over PA behaviour and were linked to previous experiences and events.
Learmonth et al. 2013	Fourteen people with Multiple Sclerosis (4 males; 10 females) who had participated in a 12 week group exercise programme specific to Multiple Sclerosis.	Multiple Sclerosis	A sem-structured focus group schedule was used to elicit participant views on the exercise class, outcomes from exercise and the exercise class and any perceived facilitators or barriers to exercise.	The condition specific, group exercise programme was enjoyed by the participants. Three themes emerged; "the exercise class" (structure, instruction, venue), the "benefits of the class, helping them to overcome" and "barriers to exercise".

O'Brien et al 2008	Participants (n=13) of a 10-week community based strengthening programme for people with Parkinson's disease as part of a research study, (3 females; 10 males).	Parkinson's disease	Individual semi-structured interviews were conducted with participants following completion of the exercise programme. Interviews explored the positive and negative aspects and outcomes of the programme, motivators to begin and continue with the programme, and facilitators and barriers to completion of the programme.	Four themes were identified from the interviews. They were "Motivators for participation in the PRST programme were broader than physical outcomes", "outcomes of the programme were broader than just physical outcomes"; people described aspects of social and psychological motivators and benefits "indicators of success for participants varied" and, "the participants' experience of a disease-specific exercise programme was positive".
Plow et al 2009	Thirteen participants (predominantly females) who were participating in a larger clinical trial with Multiple Sclerosis and classified as physically active, sometimes active or inactive.	Multiple Sclerosis	Individual semi-structured interviews were used to explore PA behaviour of persons with Multiple Sclerosis (MS), identify facilitators and barriers to PA, and explore the utility of Social Cognitive Theory (SCT) and Transactional Model of Stress and Coping (TMSC) in understanding PA behaviour in MS	Inactive and active participants differed in their self-regulation skills, self-efficacy and coping styles. Common barriers to PA included symptoms and the physical and social environment. Facilitators included strong self-regulation skills, confidence to overcome symptoms to engage in PA (i.e. barrier self-efficacy) and positive coping styles. Authors suggest that PA interventions will need to implement multiple strategies that target self-efficacy, social environment and coping styles. Social cognitive theory and the Transactional Model of Stress and Coping were useful in understanding PA behaviour among persons with Multiple Sclerosis; however, a limitation to these theories is that they are not explicit in the relationship between health and cognitions.

Ravenek et al 2009	People with early stage Parkinson's disease	Parkinson's disease	Background questionnaire and semi-structured interviews were used to investigate how social support influences PA participation, as perceived by men and women in the early stages of Parkinson's disease (PD). It will also address how this perceived support interacts with perceptions of control to influence this participation.	First study looking at factors influencing PA participation in PD. Issues related to support for exercise from physicians and organisational issues of support groups for PD are discussed.
Smith et al 2011	Eleven individuals with Multiple Sclerosis (female) who experienced fatigue and regularly participated in community-based exercise activities.	Multiple Sclerosis	Interviews were conducted explore and describe the experiences of people with MS-related fatigue, who engaged in community-based exercise activities in order to discover how fatigue influenced their exercise participation.	Seven factors were identified that were considered by participants to be important with regards to PA; a wellness philosophy (for prevention of other long term health problems), a related goal, belief that control was possible, feeling safe and supported, being able to manage limits, being satisfied with trade-offs, developing a positive definition of self. The controlling and unpredictable 'nature of the beast' described the MS related fatigue experienced.
Skår et al 2014	Individuals with Multiple Sclerosis (n=10; 7 females, 3 males) who had participated in a physiotherapy rehabilitation study (for 2 years) for people with MS with gait problems, but able to walk.	Multiple Sclerosis	Two focus groups (n=5 in each group) explored how people with multiple sclerosis experience participating in inpatient rehabilitation, and how it might provide psychosocial benefits.	Findings indicated that during the intervention participants shared coping strategies, experiences of symptoms and social stigma which lead to a sense of 'community' and mutual recognition of ability, impairment, self and identity and ultimately empowerment through use of coping strategies to counteract the social stigma they experienced.

Cedervall et al 2015	Fourteen individuals with mild Alzheimer's disease (8 females, 6 males) of varying levels of PA participation and need for support to participate in PA.	Alzheimer's disease	Individual interviews were conducted with the participants and their partners. Interviews were conducted on three occasions, one year apart as part of a larger longitudinal study investigating PA levels and capacity (Cedervall, Kilander, & Aberg, 2012; Cedervall et al., 2014). The findings presented are based on the baseline interviews.	Themes centred around the idea of using PA as a way of maintaining self-hood 'striving to be active' and a means of achieving well-being. For participants, PA appeared to be associated with a healthy life and so their routines helped them to take the focus from the illness to a more healthy self.
Malthouse and Fox 2014	Five people with Alzheimer's disease and their spouse carers.	Alzheimer's disease	Semi-structured interviews were conducted with the participants with AD and their spouse carers with the aim of improving understanding about the barriers and facilitators to PA for people with Alzheimer's disease and their spouse carers.	Overarching themes of 'self', 'others' and 'couple' were elicited and the complex interplay between those themes meant that individually tailored approaches to support people with AD and their carers to be active is needed that considers the progressive and changing nature of AD.

Appendix 6: Summaries of studies from the literature review that used theoretical models to underpin the research

<b>Study</b>	<b>Population</b>	<b>Exposure / theoretical model</b>	<b>Outcome</b>	<b>Key significant findings and relevance of the chosen model</b>
Eriksson et al 2013	Eleven individuals (6 males) with Parkinson's disease (PD) who had been regularly participating in exercise for more than 1 year and had participated in a PD specific exercise programme.	Parkinson's disease / social cognitive model	Individual interviews to explore experiences and generate understanding of the meaning of exercise for people participating in an exercise programme.	A core category and six categories were identified. The core category related to participants' efforts to maintain function and activities through being active 'keep moving to retain the healthy self'. Other categories related to other concepts considered important to start and maintain physical activity (PA) habits and included categories of "Taking rational position", "Exercising to slow progression", "Exercising to achieve well-being" and "Using exercise as coping strategy". Having goals and self-efficacy were identified as important for adherence.
Elsworth et al 2009	Twenty four people with either muscular dystrophy, multiple sclerosis, motor neurone disease or Parkinson's disease.	Neurological conditions / social cognitive model	Four condition specific focus groups with individuals with neurological conditions. Focus groups explored opinions on facilitators of PA participation. The themes that emerged from the focus groups were used to develop a postal questionnaire for people with neurological conditions (n= 80 out of 115 completed) to further elicit information regarding necessary support, barriers, and average time spent participating in PA.	Three themes were identified; 'opinions of physical activity', 'barriers to physical activity', and 'factors that would encourage increased physical activity involvement'. Swimming, stretching and walking were the most popular activities. Barriers to participation in PA included embarrassment, perceived lack of condition-specific knowledge of the fitness professionals about neurological disease and the impact of that on exercise advice. Group based exercise sessions (for specific conditions) and the presence of specifically trained staff were perceived facilitators.

Ravenek and Schneider 2009	Three women and four men with early Parkinson's disease (PD).	Parkinson's disease (early stage) / World Health Organisation's International Classification of Functioning, Disability and Health (ICF) model	A background questionnaire and semi-structured interviews were used to explore how social support influences PA participation, as perceived by individuals with early stage Parkinson's disease (PD) and how participation is influenced by the interactions of perceived support and perceptions of control.	First study looking at factors influencing PA participation in PD. Issues related to support for exercise from physicians and organisational issues of support groups for PD are discussed.
Jones et al 2008	Twenty people with Parkinson's disease (12 male, 8 female; mean age 65 years (range 50 – 80); mean disease duration 10 years (range 2.5 – 26).	Parkinson's disease / World Health Organisation's International Classification of Functioning, Disability and Health (ICF) model	Semi-structured interviews were used to explore the personal experience of everyday walking with Parkinson's disease.	Challenges and the strategies used to compensate for difficulties were identified. Findings showed that the physical and psychosocial context of everyday walking need to be accounted for by rehabilitation interventions and care giver input can be important for monitoring and supporting walking activity.
Plow et al 2009	Thirteen participants with Multiple Sclerosis (MS) from a clinical trial of two different exercise based interventions were interviewed and classified as physically active, sometimes active or inactive	Multiple Sclerosis / Transactional Model of Stress and Coping (TMSC) and social cognitive theory (SCT)	Interviews were conducted with 13 participants of a clinical trial in Multiple Sclerosis who were classified as physically active, sometimes active or inactive. Similarities and differences were explored between groups. Interviews were analysed so as to identify facilitators and barriers to PA, and explore utility of SCT and the TMSC in understanding PA behaviour among persons with MS. Using analytical induction, data was coded into pre-established categories which were constructs from SCT (i.e. environment, expectations, self-efficacy and self-	Differences were elicited between inactive and active participants in terms of their self-regulation skills, self-efficacy and coping styles. Symptoms and the physical and social environment were described as common barriers to PA. Strong self-regulation skills, confidence to overcome symptoms to engage in PA (i.e. barrier self-efficacy) and positive coping styles were found to be facilitators of PA. SCT and TMSC were found to be helpful in gaining understanding PA behaviour in MS. Plow and colleagues highlight that the theories are limited by not being explicit in the relationship between health and cognitions and suggest that future research to explore how to incorporate models of health and function into existing behaviour change theories is needed. In light of the

			regulation) and TMSC (i.e. stress appraisal and coping style).	findings, the authors suggest that multiple strategies that target self-efficacy, social environment and coping styles are needed in development and implementation of future PA interventions.
Helder et al 2002a	Individuals with a clinically confirmed diagnosis of HD (n=77)	Huntington's disease / Self-regulation model	The Illness Perceptions Questionnaire (Weinman et al. 1996) was used to assess the five components of illness perceptions described by Leventhal et al. (1984): identity, time-line, cause, consequences, and control/cure. This was to help describe illness perceptions and coping mechanisms and their role in the well-being of people with HD.	Many symptoms of HD reported, some negative consequences of the disease for their everyday lives, disease would have a long duration, not much hope for cure or improvement of symptoms, could not state with certainty whether they had control over the disease process and the main cause was considered to be genetic with stress as an additional contributing factor to their illness. No significant differences between male vs female, employed vs unemployed on the consequences scale. Differences were seen in the identity scale between those who had to give up work because of HD and those still working and there was a significant positive correlation between duration of HD and scores on the 'identity' scale and significantly negatively associated with a disease specific motor score. In terms of coping, acceptance was seen more often in HD than in 'healthy control individuals' and sought less instrumental social support, venting of fewer emotions, suppressing fewer competing activities, and adopted fewer strategies to mentally disengage from the stressful situation (HD). Illness perceptions characterised by a strong illness identity were negatively related to patient's well-being.

Helder et al 2002b	Spouses of people with Huntington's disease (n=90)	Huntington's disease / Self- regulation model	This study investigated how coping mechanisms and illness representations contribute to quality of life in spouses of people with Huntington's disease. Interviews and questionnaires were conducted at the participants' homes. Questionnaires included the Illness Perception Questionnaire, (Weinman 1996), the COPE inventory (assesses coping strategies) and quality of life using the Medical Outcome Study 36 item Short Form Health Survey (MOS SF-36) and the Sickness Impact Profile (SIP) (Bergner, Bobbitt, Carter, & Gilson, 1981).	Spouses of people with HD perceived their partner's HD as a high number of somatic symptoms (strong illness identity), being of long duration, and having negative consequences for their day to day living. They reported having little hope for cure or control of the HD symptoms that they believed were mainly caused by genetic factors.  The authors suggest that the coping strategies adopted by spouses were similar to reference participants dealing with everyday stressful situations, therefore HD does not seem to affect their quality of life. However they also point out that the measure used for illness perceptions may have been too global therefore not sensitive to capture specific contextual issues.  Devising strategies to deal with HD was positively related to spouses' role functioning. They responded to the HD by accepting the disease and the problems caused by it, took action to solve the problems encountered, and tried to develop strategies to deal with the problems at hand.
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Kaptein et al 2006	People with Huntington's disease (n=77)	Huntington's disease / Self- regulation model	The Illness Perception Questionnaire (IPQ) was used to assess HD patients' cognitive representation of their illness to investigate the contribution of illness perceptions and coping mechanisms to the explanation of well-being of patients with Huntington's disease (HD). Coping was assessed by means of the COPE, which explores problem- focused coping ('Active coping', 'Planning', 'Suppression of competing activities', 'Restraint coping', and 'Seeking social support for instrumental reasons') and emotion-focused coping ('Seeking social support for emotional reasons', 'Positive reinterpretation and growth', 'Acceptance', 'Turning to religion', and 'Denial') (Carver, Scheier, & Weintraub, 1989). Physical and psychosocial well-being was assessed using the Sickness Impact Profile (SIP) (Bergner, Bobbitt, Carter, & Gilson, 1981).	The results suggest that coping does not mediate the relationship between illness perceptions and psychosocial functioning. Contribution of illness perceptions to the well- being of people with HD was found to be significant.
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Kaptein et al 2007	People with HD (n=36) and their partners (n=19)	Huntington's disease / Self- regulation model	The Illness perception questionnaire, Unified Huntington's disease Rating scales, mini mental state questionnaire and Medical The Medical Outcome Study 36- item Short Form Health Survey were conducted with patients and their partners, separately at home to investigate whether partners hold similar views about HD, to investigate patterns between illness perceptions of patients and their partners and quality of life and to explore the cognitive factors related to Quality of Life for couples living with HD.	In general, people with HD held more positive beliefs about HD than their partners did and attributed significantly less symptoms to HD compared to their partners. People with HD believed that the disease was more controllable than what their partners perceived. Stronger beliefs in control over the illness and less serious perceived consequences of the people with HD were related to better partner quality of life. For people with HD, a less severe illness identity and longer perceived duration of the illness, less consequences, more control and less belief in treatment were related to a better quality of life. In addition, stronger beliefs of the partners in a longer duration of disease and less belief in cure through treatment were related to higher vitality scores of the people with HD. Perceptions held by the partners of identity and consequences were not significantly related to patients' vitality, social functioning or mental health ratings. Stronger beliefs in control over HD from the people with HD were linked to higher vitality scores of partners. Spousal illness perceptions were related to vitality scores of quality of life for both people with HD and their partners, although their own perceptions were responsible for the largest variance in quality of life.
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Arran et al 2014	People with HD (n=87) completed the questionnaires online or by post (female n=49; male n= 38)	Huntington's disease / Self-regulation model	The Illness Perceptions Questionnaire was used to explore the biopsychosocial factors related to psychological distress in people with HD, and examine the relationship between illness perceptions, coping and psychological distress. Self-report measures were also completed by participants related to coping and psychological distress (SF-36 health related quality of life questionnaire and Hospital Anxiety and Depression Scale (HADS)) and data with regard to clinical and demographic variables were collected that are associated with psychological distress.	The authors found that illness representations reflect distinct and meaningful constructs for people with HD. In relation to the illness representation domain of identity, fatigue (reported by 65.5%), sleep disturbances (64.4%) and loss of strength (62.1%) were most commonly reported. Most commonly reported HD specific symptoms were involuntary movements (82.8%), problems with balance (80.5%), problems with concentration (79.3%), memory difficulties (72.4%), problems with walking (71.3%), difficulties reading and writing (69%) and personality changes (60.9%). Participants perceived HD to be chronic, as opposed to cyclical and to have strong, negative consequences. Scores on the control domains (personal and treatment) showed that participants perceived little perceived control over the HD. The SF-36 results indicated perceptions of limitations in performing all physical activities, physical health causing a high number of problems with work or other daily activities greater pain in comparison with the general population. The HADS indicated that approximately 48% of individuals possibly had anxiety (n=42) and 40% possibly had depression.
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Clare and Harman 2006	Four males and 5 females with a diagnosis of early stage dementia (Alzheimer's disease n=5; frontotemporal dementia n=2; vascular dementia n=1, dementia n=1)	Early stage dementia / Self-regulation model	Individual semi structured interviews conducted with individuals with early stage dementia to identify key elements of participant understanding and experience of dementia and a secondary stage of analysis to conduct a separate theory-driven content analysis with the aim of identifying all instances of the components of the SRM within the accounts given by the participants.	The findings highlighted representations of timeline, consequences, and controllability elements of the SRM. Problems with memory, organizational skills, and coordination were identified as symptoms, and a major concern was withdrawal into increasing isolation in the future, and uncertainty as to exactly how things would develop. The subtheme depicting the emergence of making comparisons suggests that participants were using a wide range of information to construct their representations which seemed closely linked to participants' efforts at coping. Participants identified ways of trying to manage the condition and deal with the challenges it posed to their sense of identity and personal integrity. Participants recognised that not all coping strategies were successful, often needing ongoing adjustment. Illness representations and the SRM are useful frameworks to help develop understanding of how people with early stage dementia attempt to manage the threats to self that onset of the condition imposes as they deal with it on a daily basis. The illness representations reflect understanding of a progressive decline linked with the significant concrete consequences that create a significant challenge.
Matchwick et al 2014	Four men and 2 women with Alzheimer's disease with an age range of 71 to 83, (mean age of 77)	Alzheimer's disease / Self-regulation model	Individual semi-structured interviews explored the cause and control domains of illness representations in older adults with Alzheimer's disease (AD).	Findings identified three main themes. These gave indication that participants tried to make sense of their Alzheimer's disease by comparing it with previous experiences of physical illnesses. Participants engaged with their diagnosis in a graded way because of the lack of tangible diagnostic evidence and developed pragmatic emotional responses to their situation.

## Appendix 7: ENGAGE-HD: summary of researcher's involvement, workshop schedule and intervention development paper

As part of the researcher's job role as research assistant working with Professor Monica Busse, in 2013, the researcher was to support delivery of workshops around the UK with people with HD, family members and caregivers. The purpose of the workshops was to gain feedback about a 'physical activity workbook' that was being developed for use in the ENGAGE-HD trial to support those participants randomised to the physical activity intervention group and also to help design the physical activity intervention itself. Around the same time, the researcher had developed ideas about their own research as described in the section 'background of the researcher'. It was therefore negotiated and planned with Prof Busse that the researcher would have time during the workshop days to conduct their own qualitative research. To address the aims of the researcher's research, people across the spectrum of HD and caregivers/family members were required and so the invitations and information sheets enabled inclusivity of all stages as long as participants could take part in discussions in English. As described in the methods section, the researcher decided that focus groups were the most appropriate data collection method to address the aims of their research and so during the workshops, the researcher conducted focus groups with participants (described in 'methods').

The workshop days were split into two parts; the researcher's specifically developed focus groups were conducted first (schedule can be seen in appendix 13), followed by the workshops/focus groups for ENGAGE-HD. The schedule for the ENGAGE-HD session can be seen below.

1. We received feedback from a series of questionnaires given to people with HD, their family members and carers and HDA care advisors. These people have said that they need more information about exercise in HD. Do you agree with this? Please tell us more.

**PROMPTS**

*What would be helpful?*

*In what form would you like more information?*

2. What specific advice is important for you know about exercise?

**PROMPTS**

*How do I know how often I should exercise?*

*How do I know which exercises are best (and safe) to do?*

*How do I decide where the best place for me to exercise is? [This may relate to environment/weather/home safety]*

*How do I know how hard I should push myself? How do I know when I have done enough exercise? How do I know when to do more exercise?*

*Will exercise make me more tired?*

3. Our studies suggest that people who have a willing family member or carer to support them are more able to exercise regularly. What do you think about this? What are the implications for the family member or carer?

**PROMPTS**

*Is this a true reflection of the reasons why people may adhere better to an exercise programme or are there other important factors?*

*Does care burden increase if supporting exercise for the person with HD? If so, are there longer term benefits to be gained by supporting the person with HD to exercise? Does the potential for longer term benefits outweigh the commitment required?*

4. "In the past week/past month, on how many days have you done a total of 30 minutes or more of physical activity, which was enough to raise your breathing rate. This may include sport, exercise, and brisk walking or cycling for recreation or to get to and from places, but should not include housework or physical activity that may be part of your job".

5. We would like to show some questionnaires and an example workbook and ask you thoughts about this?

Give a summary of the discussion after Part 2 and ask "Is this an adequate summary?"

"This is the first in a number of groups like this that we are organising, would you be happy for us to contact you to talk about your experiences shared in this discussion after this meeting to confirm we have the correct information?"

# Development and Delivery of a Physical Activity Intervention for People With Huntington Disease: Facilitating Translation to Clinical Practice

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**Background and Purpose:** We studied the development and delivery of a 14-week complex physical activity intervention for people with Huntington disease, where detailed information about the intervention was fully embedded in the trial design process.

**Methods:** *Intervention Development:* The intervention was developed through a series of focus groups. The findings from the focus groups informed the development of a logic model for the physical activity intervention that was broadly consistent with the framework of self-determination theory. *Intervention Delivery:* Key components underpinning the delivery of the intervention were implemented including a defined coach training program and intervention fidelity assessment methods. Training of coaches (physical therapists, occupational therapists, research nurses, and exercise trainers) was delivered via group and 1:1 training sessions using a detailed coach's manual, and with ongoing support via video calls, and e-mail communication as needed. Detailed documentation was provided to determine costs of intervention development and coach training.

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**Results:** Intervention delivery coaches at 8 sites across the United Kingdom participated in the face-to-face training. Self-report checklists completed by each of the coaches indicated that all components of the intervention were delivered in accordance with the protocol. Mean (standard deviation) intervention fidelity scores ( $n = 15$ ), as measured using a purpose-developed rating scale, was 11 (2.4) (out of 16 possible points). Coaches' perceptions of intervention fidelity were similarly high. The total cost of developing the intervention and providing training was £30,773 (\$47,042 USD).

**Discussion and Conclusions:** An important consideration in promoting translation of clinical research into practice is the ability to convey the detailed components of how the intervention was delivered to facilitate replication if the results are favorable. This report presents an illustrative example of a physical activity intervention, including the development and the training required to deliver it. This approach has the potential to facilitate reproducibility, evidence synthesis, and implementation in clinical practice.

**Video Abstract** available for more insights from the authors (see Supplemental Digital Content 1, <http://links.lww.com/JNPT/A122>).

**Key words:** complex interventions, fidelity, Huntington disease, logic models, physical activity intervention development

(JNPT 2016;40: 71–80)

## INTRODUCTION

Huntington disease (HD) is a dominantly inherited neurodegenerative condition that affects the brain, causing dysfunction and death of medium spiny striatal projection neurons and thus disruption of corticostriatal pathways, with resultant gradual impairment of cognition and motor function, along with behavioral problems including apathy, anxiety, and irritability.<sup>1</sup> Currently there is no disease-modifying treatment available for this condition, and very little known regarding effective symptomatic treatment. Encouraging regular physical activity throughout the developing disease may offer a means to enrich the lives of people with HD and their carers by helping to maintain independence, improve health, and subsequently reduce health and care costs.

The benefits of physical activity in maintaining cardiovascular health and reducing mortality in the general population are widely recognised,<sup>2</sup> and there is an ever-increasing

public health focus on physical activity for maintenance of health. Exercise interventions also seem to have similar, if not potentially better, mortality outcomes among a range of chronic diseases compared with drug interventions.<sup>3</sup> There is also a growing interest in the potential of regular physical activity in people with neurodegenerative conditions, such as multiple sclerosis, Parkinson disease, and Huntington disease. Exercise and physical activity are secondary prevention strategies that have the potential to significantly impact the progression and management of neurodegenerative diseases, including maintaining function and improving postural control, gait, and health-related quality of life.<sup>4–6</sup> However, many healthy individuals and those with neurodegenerative diseases have difficulty maintaining adherence to exercise programs. Developing interventions that are aimed at specifically promoting adherence and facilitating exercise uptake have thus been the focus of emerging research.

One of the challenging aspects of this developing research is achieving effective translation from research to practice. Even if studies demonstrate positive effects, implementation may not readily occur. One of the possible contributing factors to this may be the lack of detailed description of the various components of physical therapy interventions, which are often complex in nature. Guidelines for reporting interventions stress the importance of having well-defined, detailed descriptions of intervention components, including duration, dose or intensity, mode of delivery, essential processes, and a means of monitoring fidelity.<sup>7</sup> Furthermore, elements of the intervention should have explicit descriptions of theoretical foundations. It is encouraging that there has been a gradual increase in research focusing on understanding the components of physical activity interventions in neurologic diseases. For example, a series of theory-based interventions underpinned by established associations between social cognitive theory (SCT) constructs and physical activity have been developed for people with multiple sclerosis.<sup>8</sup> These interventions aimed to support behavior change through focusing on participants' self-efficacy, goal-setting, and outcome expectations.<sup>9–11</sup> The Blue Prescription intervention has been implemented for people with multiple sclerosis in New Zealand, with a focus on combining professional help with self-help to increase physical activity.<sup>12</sup> This study was underpinned by concepts related to motivational interviewing and promoting self-efficacy. In the Netherlands, Van Nimwegen and colleagues<sup>13</sup> developed a physical therapy intervention for patients with Parkinson disease called ParkFit, which was also explicitly based on behavior change theories, such as SCT and the transtheoretical model of health behavior change.

Although theoretical frameworks do provide some support for the interventions mentioned previously, there is a lack of consistent linkage of these frameworks within the evaluation of such interventions to inform implementation into clinical practice. For example, logic models, which graphically depict the proposed relationship between activities and expected outcomes,<sup>14</sup> are not routinely described and many studies in neurologic physical therapy, even if the intervention is described in detail, do not extend the approach to explicitly measure whether the intervention was delivered as it was

intended (ie, fidelity). An additional challenge in designing physical activity interventions for patients with neurodegenerative diseases is the need to ensure that any theoretical framework is grounded in and relevant to the particular experiences and needs of the specific population. Given that these complex diseases require a high degree of care over the disease trajectory, it is particularly important to understand and account for the views of patients, families, and carers so as to make the intervention acceptable to the intended population.<sup>15</sup>

The purpose of this report is to describe the development and delivery of ENGAGE-HD, a single-blind, exploratory phase II multisite randomized, controlled trial of a 14-week physical activity intervention compared with a social contact control intervention (ISRCTN65378754).<sup>16</sup> Multicenter research ethical approval was granted by South East Wales Research Ethics Committee B (approval number: 14/WA/0034).

Forty-six participants with genetically confirmed HD were recruited to the study; twenty-two participants were allocated to the physical activity intervention; 6 participants in this arm were withdrawn, and a total of 16 completed the intervention. The physical activity intervention involved 6 home visits from activity coaches, delivered over 14 weeks, with interim supporting phone calls. Although the protocol for this study has recently been published,<sup>16</sup> this report did not provide aspects that might facilitate successful clinical implementation of the intervention. Here, we present details of the study that are essential to promote effective knowledge translation, with consideration of user perspectives, incorporation of a theoretically grounded logic model, coach training program, fidelity methods, and costs of intervention development and delivery.

## METHODS

### Development of the ENGAGE-HD Intervention

The ENGAGE-HD intervention is grounded in an established behavioral change theory, chosen because it was judged (by analyses of focus group results described below) to be the most appropriate for the complex needs of this population. A structured logic model then guided intervention delivery, and there was a system in place for promoting and evaluating therapist fidelity. Each of these unique features, which we argue should be more widely utilized in design of clinical trials, particularly in patients with complex health conditions such as neurodegenerative diseases, will ultimately help to facilitate translation of the results from this randomized trial into clinical practice.

### Focus Groups

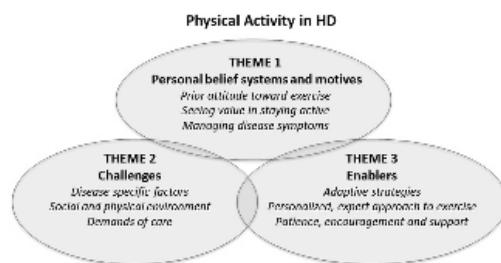
The underlying theoretical framework for the ENGAGE-HD intervention was developed through a series of focus groups. A purposive maximum variation sampling approach<sup>17</sup> was used to capture varied perspectives from people with HD, their family members, carers, and professionals. People with HD and their caregivers (both formal carers and informal carers, ie, family members) were invited by post via regional care advisors of the Huntington's Disease Association of England and Wales (HDA). The HDA maintains a confidential mailing list of members who have agreed to be contacted in this way.

All correspondence was initiated by the HDA, and no personally identifiable details were provided to the research team without the consent of the involved individuals. Eight focus groups including a total of 56 people were conducted. Of these, 26 were people with HD (46.4%; 18 male), 24 were carers or family members (42.9%; 18 female), and 6 were professionals (10.7%; 2 physical therapists, a physical therapy assistant, a health care assistant, an occupational therapist, and a nurse). The number of participants in each group ranged between 3 and 12. Several participants were at an early stage of disease progression and still able to live relatively independently; one participant was gene positive but asymptomatic. Others were at a much later stage of the disease and more severely disabled by their symptoms. Across all participants, there was involvement in a variety of activities ranging from relatively low intensity such as walking or gardening to more vigorous exercise such as running.

Focus group facilitators were all registered physical therapists with experience working with people with HD and their families. A single facilitator moderated each focus group using a semistructured topic guide covering 4 key areas about the physical activity experiences of people with HD: (1) descriptions of these experiences; (2) impact of the disease; (3) carer's experience; and (4) clarifying enablers for regular physical activity. When moderating the group, special attention was given to the needs of people with HD and the role of the family members and carers. Reframing, repetition, and expansion of the questions as required were used to encourage full participation of all present. A second facilitator was also present in each group to capture field notes. HDA care advisors were also in attendance at all meetings. In 3 of the 5 locations (Cardiff, Southampton, and Liverpool), 2 focus groups were conducted in parallel. Focus groups were digitally audio-recorded and transcribed verbatim; the accuracy of the transcripts was confirmed by both the focus group leader and the field notes.

Focus group audio-recordings and transcripts were analyzed thematically.<sup>18</sup> These themes were identified as patterns in the discourse of focus group participants that corresponded with the research questions. The coding frame was developed inductively through an iterative process of data analysis. A second researcher double coded 25% of the data (2 of the 8 focus groups). Where 95% agreement was reached between the 2 coders, no action was taken. Alternately the coders reviewed areas of discrepancy and resolved these. There were no coding discrepancies that could not be resolved. QSR NVivo10 software was used (QSR International Pty Ltd 2014).

Three themes emerged from the focus groups: personal beliefs and motives, enablers, and challenges (Figure 1). Although many of the enablers and challenges to physical activity were not unique to this population—the challenge of integrating physical activity into existing schedules, for instance—other disease-specific factors were highlighted. In particular, it was clear that people with HD wanted to be provided with a range of options and be able to choose what sort of activities they might take part in, rather than being prescribed a rigid exercise plan. Although some patients enjoyed the social aspect of group activities such as golf, for instance, most were reluctant to exercise in public because of what they perceived to be a social stigma attached to their disease. Likewise, carers



**Figure 1.** The 3 themes that emerged from the focus groups: personal beliefs and motives, enablers, and challenges.

spoke of the need to tailor activity plans to the ability of the person with HD, rather than seeing each person as "just an HD sufferer." For many patients whose activity levels were very limited, family members and carers suggested that starting with simple activities such as getting out of a chair without any assistance would allow people with HD to gradually build confidence. Patients themselves spoke of wanting specialist support and advice, to help them find activities that might be suitable for their condition. Finally, carers highlighted the need for patience, encouragement, and empathy when working with individuals with HD, in order to slowly build trust and help patients overcome the fear of falls or experiencing pain when exercising.

The findings from the focus group were interpreted as being broadly consistent with the framework of self-determination theory (SDT).<sup>19</sup> Self-determination theory is a theory of human motivation that has been applied across a range of health behaviors, including physical activity. Self-determination theory suggests that motivation in general, and indeed with respect to physical activity, can be placed along a continuum from extrinsically motivated and regulated (for rewards or to satisfy an external demand) to the more autonomous, intrinsically integrated, and self-determined behavior as the motives become internalized. Self-determination is said to arise from feelings of autonomy (being in control of behavior and having choices), competence (experiencing a sense of mastery or skill), and relatedness (feeling connected to and understood by others).

Our participants described a range of regulatory styles along a continuum from intrinsic to extrinsic that could potentially have an impact on sustained physical activity behaviors. Some participants talked about physical activity as enjoyable and essential to their quality of life, and others participated only with sufficient encouragement from carers. In their talk of wanting to maximize independence, to challenge themselves and to improve their health, people with HD described intrinsic goals, which are associated with enhanced participation in exercise.<sup>19,20</sup> However, these participants also experienced considerable challenges through their HD symptoms, such as loss of insight, balance, and motor function, which could negatively impact their ability to safely perform physical activity. Much like interventions that have incorporated a SDT framework with a psychiatric

population, where motivational mechanisms were not different from those in the normal population even in the presence of disease-specific barriers to physical activity,<sup>21</sup> we suggest the motivational processes underlying physical activity behavior in people with HD may be at least partially explained using this theory.

### Development of Logic Model

This complex intervention consisted of 3 main elements, namely the *participant/coach interaction* (underpinned by SDT), a purpose developed *ENGAGE-HD Workbook*, and an exercise DVD (*Move to Exercise*).<sup>6,22</sup> Each of these elements is described in detail below. Figure 2 presents this in the form of a logic model, describing the key elements (inputs) and activities of the physical activity intervention (outputs).

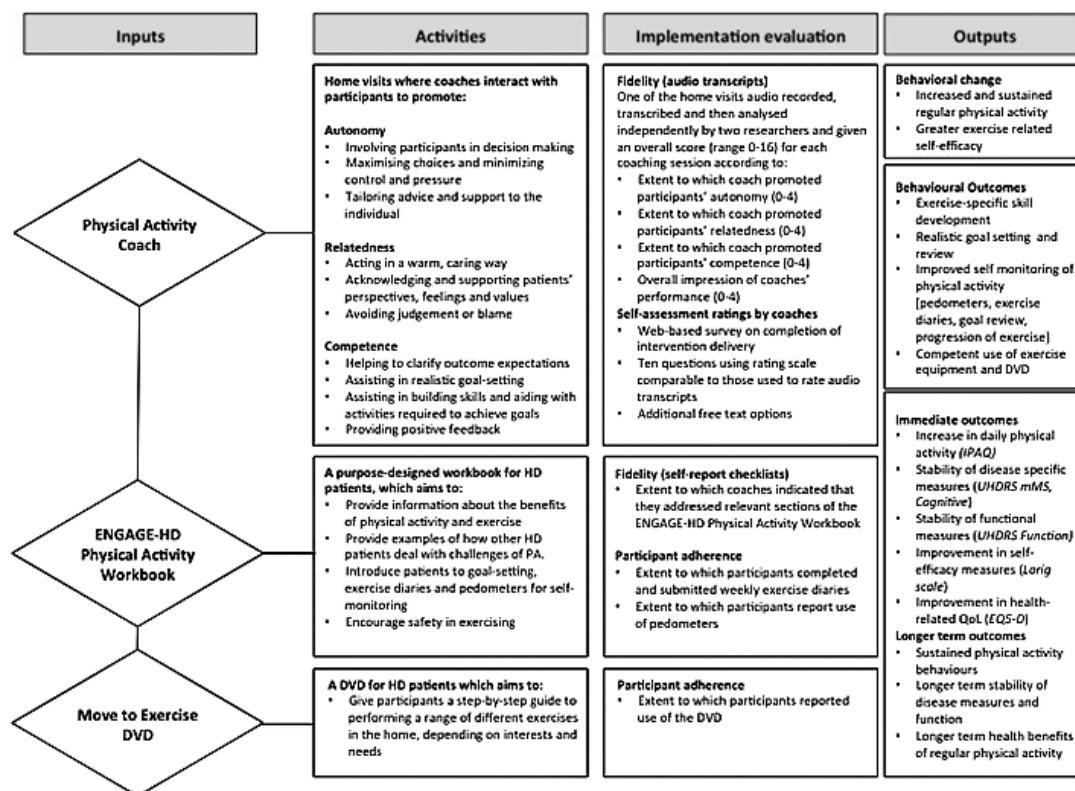
### Participant/Coach Interaction

The coaching visits and the participant/coach interaction in the *ENGAGE-HD* intervention specifically aimed to develop self-determined physical activity behaviors through in-

tentionally promoting feelings of autonomy, competence, and relatedness. Specifically, coaches were encouraged to promote *autonomy* by involving participants in any decisions, minimizing control or pressure, and tailoring advice and support to the individual. *Competence* was promoted through helping patients to clarify potential outcomes of physical activity, working with participants to set realistic and measurable goals, and providing positive feedback. Finally, *relatedness* was promoted through acting in a warm and caring way, expressing empathy and avoiding judgment and blame.

### ENGAGE-HD Workbook

The *ENGAGE-HD* workbook was used as a guide for the interactions. Workbook-based approaches have been used to promote self-management approaches in other diseases and disorders, including the Bridges program used in patients post-stroke.<sup>23</sup> During the first home visit, the coach introduced the participant to the program and the workbook. The workbook is structured into 5 distinct sections: (1) Exercise-Who Me?; (2) HD Experiences in Increasing Physical Activity; (3) Goals; (4) My Physical Activity Plan; and (5) Recording Progress.



**Figure 2.** The logic model describing inputs, activities, and outputs. This has been adapted from the *ENGAGE-HD* trial protocol paper (open access article distributed under the terms of the Creative Commons Attribution License, <http://creativecommons.org/licenses/by/4.0/>,<sup>16</sup> to provide additional detail with respect to methods for implementation evaluation).

The initial interactions consider benefits of physical activity and each participant's individual exercise history. Participants are encouraged to identify specific areas in their lives (both formal and informal) that could be altered to promote physical activity for general well-being, and also to set specific physical activity goals. Instructions were provided for assuring safety of exercise, including use of perceived exercise scales, and also for use of pedometers (provided to participants) to measure physical activity. Further discussion topics on physical activity include implementing a daily activity plan, monitoring exercise intensity, and dealing with safety, weather, equipment, and typical barriers (eg, time, boredom, lack of equipment, lack of specific knowledge, and support).

### Move to Exercise DVD

The final component of the ENGAGE-HD intervention is the *Move to Exercise* DVD. The *Move to Exercise* DVD was developed after consultation with people with HD, their family members, and physical therapists specializing in neurodegenerative diseases,<sup>24</sup> and has been found to be acceptable and of benefit to people with HD.<sup>6,22</sup> The individual DVD sections can be used differentially based on an individual's specific needs and targets, and the coaches work with the participants to identify relevant sections that may be appropriate for them. Although the exercise DVD is optional, it provides a specific activity, in addition to walking programs, that the coaches can focus on to facilitate increased physical activity.

### Delivery of the ENGAGE-HD Intervention

An intervention in a clinical trial must be delivered in a systematic manner to facilitate translation of the intervention into clinical practice, if the results of the trial indicate it is safe and has potential for benefit. Key components of effective delivery are training of the coaches delivering the intervention, providing ongoing support, assessing costs for training and support, and fidelity monitoring.

### Training of Coaches

The coaches delivering the ENGAGE-HD interventions were either (a) health care professionals (eg, physical therapists, occupational therapists, or nurses) with experience

of delivering exercise-related activities or with specific experience with HD; or (b) exercise professionals. All staff had to meet specific health competencies, namely Skills for Life Competencies, developed by the National Health System (NHS) in the UK. (Competencies can be found at Skills for Life, accessed May 29, 2015: <https://tools.skillsforhealth.org.uk/competence/show/html/id/2603/>). Nevertheless, across the sites, the coaches would likely have a wide range of backgrounds and experiences, hence the need for centralized and standardized training and support.

The training model was for a team, including the intervention coordinator, trial chief investigator, and trial manager to travel to the site location and conduct a 6-hour training session in a small group setting (Table 1 for overview of the training program). Training for the coaches included a 1.5-hour, one-to-one session with either the chief investigator or the intervention coordinator. Both the chief investigator and the intervention coordinator were research physical therapists with extensive experience working with the HD community in both clinical practice and research, who oversaw development of the training materials and ongoing support of the coaching staff. A coach's manual was provided to each coach, and was used as a guide for each of the training sessions. The coaching manual gave an explicit, session-by-session guide, familiarized the coaches with the specific challenges of working with patients with HD, and offered a background to the intervention's SDT framework.

### Ongoing Support and Monitoring

In addition to the initial training sessions and coaching manuals, coaches received ongoing support from the intervention coordinator. This support is particularly important in helping to guide coaches who have had little or no experience of working with patients with this relatively rare disease. Before each coach visited a participant for the first time, they were able to have a discussion with the intervention coordinator to assist them to interpret a participant's baseline assessment scores (including measures of walking ability, cognitive function, a motor score, and a breakdown of scores on the Physical Performance Test). This allowed them to appropriately anticipate the ability level and potential needs of each participant. After the initial home visits, coaches

**Table 1. Summary of Training, Support, and Monitoring for Physical Activity Coaches**

	Time	Description
Initial training	6-h training session for all site staff, including coaches with individualized 1.5-h training	<ul style="list-style-type: none"> <li>• Delivered by either trial PI or the intervention coordinator</li> <li>• Review of coach's manual, with explicit, session-by-session guide</li> <li>• Familiarized the coaches with the specific challenges of working with patients with HD</li> <li>• Offered a background to the intervention's SDT framework</li> <li>• Before first visit, coaches had video discussion with the intervention coordinator to assist them in interpreting a participant's baseline assessment scores</li> <li>• After the first or second home visit, coaches had a further discussion with the intervention coordinator to discuss goal setting and address any concerns or issues</li> <li>• Coaches were further encouraged to contact the intervention coordinator if they had any questions about the home visits as the intervention progressed, either by e-mail or videoconferencing</li> </ul>
Ongoing support	Minimum 2 discussions, others as needed	

Abbreviations: HD, Huntington disease; PI, principal investigator; SDT, self-determination theory.

had a further discussion with the intervention coordinator to develop realistic goals for the participants, based on each participant's particular interests and their current ability levels. Coaches were further encouraged to contact the intervention coordinator if they had any questions about the home visits as the intervention progresses, either by e-mail or video-conferencing.

### Fidelity Monitoring

Fidelity of an intervention measures the extent to which the intervention was delivered in the way it was intended. In this study, fidelity was measured for each of the 3 elements of the intervention: the coach interactions, the Physical Activity Workbook, and the Move to Exercise DVD. Fidelity was measured by a combination of self-report checklists, independent analysis of audio-recordings, and a self-assessment completed by the intervention coaches.

After each of the 6 home visits, coaches were required to complete a short self-report checklist, indicating whether the content of each of the sessions was consistent with what was specified in the protocol and training manual. For visit 1, for instance, the checklist asks whether the coach introduced the ENGAGE-HD program, talked to the participant about the exercise workbook and DVD, and whether he or she discussed the idea of setting a series of activity-based goals. The checklists also recorded the number of minutes that coaches spent delivering each session.

Recognizing the limitation of self-report measures of intervention fidelity, we also included an independent assessment of the quality of the coaching sessions, based on audio-recordings of one of the coach home visits. The fidelity of the coach interactions was measured by assessing the extent to which each coach demonstrated efforts to promote a patient's autonomy, relatedness, and competence. Coaches were asked to audio-record one of their later home visits (typically the

third of 6 visits). The audio files were transcribed and then independently rated by a member of the study team, using a rating scale that represented the core features of the intervention as described in the logic model (Table 2). For rating on the scale, coaches were given a 0 to 4 rating for the 3 SDT areas (autonomy, competence, and relatedness) and a final 0 to 4 score to reflect an overall impression of the coach's performance. Each recorded coaching session was accordingly scored from 0 to 16. Scoring of the sessions using this rating scale had 2 purposes: (1) the lead intervention coordinator was able to use the transcripts to provide coaches with constructive feedback on their interactions in-between visits, promoting ongoing fidelity; and (2) individual fidelity scores could be used as a potential mediating factor when exploring measures of benefit (blinded outcome measures). To ensure that the fidelity rating tool could be readily implemented in a clinical setting utilizing relatively novice raters, the study team member (who was a researcher and not involved in delivery of the intervention) and the intervention coordinator independently rated 3 audio files and compared ratings for agreement. For each of the 5 possible levels within each of the 4 items (autonomy, competence, relatedness, and overall impression), the ratings for the 2 raters were within one point of each other, and for 2 of the 3 total scores, there was 100% agreement.

Fidelity of the intervention was further evaluated by asking coaches to complete a self-assessment of their perceived ability to deliver the intervention as it was intended to be delivered. We surveyed those coaches that had delivered the intervention for their opinions on the content and structure of the intervention and the issues surrounding its delivery. A set of 10 questions with a mix of rating scales (directly comparable to those scores used to rate fidelity) and free text answers were developed and delivered to the coaches via a web-based survey. The questions covered each coach's views on the training provided (including the audio-recording of one visit to assess

**Table 2. Rating Tool Used to Assess Fidelity of Delivering the ENGAGE-HD Intervention**

Item	Description	Score				
1. Autonomy	<ul style="list-style-type: none"> <li>• Involves participants in decision making</li> <li>• Minimizes control and pressure</li> <li>• Maximizes participants' choices</li> <li>• Provides a rationale for suggestions</li> <li>• Allows the participant to overtly express the pros and cons of changing behavior</li> <li>• Tailors advice and support</li> </ul>	Not at all 0	1	2	3	A great extent 4
2. Relatedness	<ul style="list-style-type: none"> <li>• Acts in a warm and caring way</li> <li>• Expresses empathy</li> <li>• Acknowledges and supports patients' perspectives, feelings, and values</li> <li>• Avoids judgment or blame</li> </ul>	Not at all 0	1	2	3	A great extent 4
3. Competence	<ul style="list-style-type: none"> <li>• Helps to clarify outcome expectations (what a person might expect as result of the changes that they have made)</li> <li>• Assists in realistic goal-setting and developing a tailored activity plan</li> <li>• Assists in building skills and developing coping strategies required to achieve specific goals</li> <li>• Provides positive feedback</li> </ul>	Not at all 0	1	2	3	A great extent 4
4. General impression	<ul style="list-style-type: none"> <li>• Overall perception of participant/coach interaction is positive</li> <li>• Coach is in command of the session and demonstrates ability to direct conversation and maintain focus</li> </ul>	Not at all 0	1	2	3	A great extent 4

fidelity), adherence of the intervention to SDT, accompanying materials used in the delivery of the intervention, and the intervention in general. Respondents were asked to identify themselves so that their answers could be linked to individual fidelity scores.

### Costs of Intervention Development and Training

The costs of developing the intervention included costs of conducting the focus groups, encompassing recruitment material, venue hire and refreshments, travel reimbursement for staff and participants, staff time attending the focus groups and interpreting output, and transcription costs. These costs also included the costs of developing the workbook and the exercise DVD. This included staff time to develop the content, design fees for the workbook and the DVD, and licensing fees for the DVD.

A spreadsheet was used to record the travel and subsistence costs for the training team, the number of hours spent travelling to training, the number of hours spent in training (for both the training team and staff being trained), venue hire, and refreshment costs. The midpoint of the pay grade for each staff member attending training was used to calculate the hourly cost of their time, including UK National Insurance and pension on-costs. The cost of training varied by site and was largely influenced by travel and subsistence costs, reflecting the distance of the intervention site from the training team's base in Cardiff, UK.

## RESULTS

### Training the Coaches

Intervention delivery coaches were trained at a total of 8 sites. Coaches were a mixture of research nurses, physical therapists, occupational therapists, clinical researchers, and exercises trainers/scientists (Table 3). Almost all of the coaches had some experience with working with patients with neurodegenerative diseases, and many had direct experience in working with patients with HD.

Over the course of the recruitment period, it became necessary to train additional staff for some sites because of staffing issues. These additional staff received telephone, web-based, or face-to-face training either individually or in pairs.

In addition to the site training, the intervention coordinator provided ongoing coaching and training for the physical activity coaches. The intervention coordinator had a minimum of 2 additional contacts with the coach per participant, which were carried out over web-based video conferencing or phone. In addition to these set contact times, there was frequent e-mail communication and occasional additional video coaching as needed (a range of 1-4 additional contacts, including e-mail and video conferencing). During these sessions, the intervention coordinator was able to provide detailed advice and guidance to assure the coaches provided the intervention as intended, and further to provide advice and support for any HD-specific issues. The coordinator documented all contact.

### Challenges to Delivery of Intervention

The most notable challenge of delivering the intervention was training and support of the coaches, who had a wide range of health professional backgrounds and experiences. Although all staff met the defined competencies, there were some staff with fairly limited experience in delivering physical activity interventions, thus requiring greater initial contact and support. Disease-specific issues also needed consideration in planning the intervention delivery for all of the coaches. Coaches needed to be considerate of patient's individual schedules and preferences for appointment times, and often needed to work closely with family members and carers. In all cases, family members were integral to the intervention delivery, both to schedule and to facilitate uptake of the physical activity program. Some participants struggled with formulating physical activity goals, HD-specific concerns, such as apathy and behavioral concerns, also resulted in an increased need for support and advice from the intervention coordinator.

### Fidelity of the Intervention

Seventeen participants completed physical activity interventions, which were delivered by 7 coaches (Table 3). The self-report checklists completed by each of the coaches at the first home visit 1 indicate that in 100% of sessions (16/16), coaches introduced the participants to the Physical Activity Workbook, gave the participants the exercise DVD and discussed the concept of goal-setting with the participant

**Table 3. Qualifications and Backgrounds of Physical Activity Coaches**

Coach #	Qualifications/Background	Number of Participants in Physical Activity Intervention	Experience of Working on Physical Therapy Interventions	Experience of Working With Patients With Neurologic Conditions	Experience of Working With Patients With HD
1	Research nurse, health visitor	5	No	Yes	Yes
2	Research nurse	3	No	Yes	No
3	Physical therapist	3	Yes	Yes	Yes
4	Occupational therapist	2	Yes	Yes	Yes
5	Research nurse	2	No	Yes	No
6	Exercise instructor	1	Yes	Yes	Yes
7	Exercise scientist, neurovascular researcher	1	Yes	No	No

Abbreviation: HD, Huntington disease.

in 100% of the sessions (16/16). Sessions lasted on average 72.3 minutes.

Fidelity scores for coach interactions, based on audio transcripts of the third intervention session, were assessed for 15 of the 16 participants. Overall scores ranged from 7 to 14 out of a possible 16 points, with a mean (standard deviation) score across the coaches of 11.0 (2.4). Coach interactions scored an average of 2.5/4 for autonomy, 3.0/4 for relatedness, 2.7/4 for competence, and 2.8/4 for the overall impression.

All 7 of the coaches completed self-assessment surveys pertaining to intervention fidelity. Self-assessment scores were on average higher than those assigned by the independent rater, namely 3.1/4 for autonomy, 3.3/4 for relatedness, and 3.0/4 for competence. In relation to the process of audio-recording a session, one coach reported that they found it "distracting" and another reported that the process may have influenced their behavior as they were acutely more aware of asking open questions during the session. Only one coach reported that the recording of the session may have affected participants adversely, making the discussion less free than it might otherwise have been. For the remainder of the coaches, they reported no difficulties or undue influences from recording the session.

Three coaches reported perceived barriers to delivering the intervention. Generally, these were logistical issues; the difficulty of scheduling home visits as per protocol in conjunction with other commitments (both for the coaches and participants) or when there had been a change in the participant's home life or disease state. One coach (a research nurse) responded that lack of confidence may have prevented the coach from being as assertive as was perhaps needed.

### Costs of Intervention Development and Training

The total cost of developing the intervention was £30,773 (\$47,042 USD). This included the cost of developing the workbook, developing and producing the DVD, and conducting the 5 focus groups. The total cost for delivering training at all of the sites, and for the additional staff training throughout the trial to date, was £18,821 (\$28,771). Costs for delivering the intervention are not reported here, and will be reported with the main study findings.

### DISCUSSION

This report describes the approach used for the delivery of a trial of a complex intervention in people with a neurodegenerative disease; the intent of the approach was to seamlessly ensure the implementation of research to clinical practice. Therapist-led interventions aimed at increasing patients' physical activity require an interaction between therapist and patient, and can typically be considered complex interventions; that is, interventions involving many interactive components.<sup>25</sup> Importantly, the theoretical basis for the complex intervention should be explicitly defined,<sup>26</sup> a recommendation that has been echoed by researchers evaluating physical therapy interventions for patients with neurologic disorders.<sup>27</sup>

An important consideration in promoting translation of clinical research, such as the intervention presented here, is

the ability to convey the detailed components of how the intervention was delivered to facilitate replication if the results are favorable. In this report, we present a detailed description of a physical activity intervention as an illustrative example, including the development and the training required to deliver it. This approach has the potential to facilitate reproducibility, evidence synthesis, and implementation in clinical practice. Additional details pertaining to the design of the study, including assessments, the control group, and additional details of intervention delivery have been previously reported.<sup>16</sup>

The ENGAGE-HD intervention included a theoretically grounded logic model, where components of the intervention were defined to inform evaluation. Crucially, the concepts related to the theoretical framework have been integrated throughout all aspects of this randomized controlled trial design: during the development of the intervention, its delivery, and its evaluation. We argue that this approach is essential to ensure knowledge translation to clinical practice. The intervention reporting is consistent with TIDieR guidelines,<sup>7</sup> and was developed in line with the Medical Research Council of the United Kingdom's Framework for Development and Evaluation of Complex interventions (MRC framework).<sup>26</sup> The MRC framework advocates the use of a cyclical development process, whereby all the components are fully developed and evaluated in an iterative process so as to ultimately ensure widespread and sustainable implementation of a specific intervention.

It is useful to inform the understanding of the components and mechanisms of the intervention to make inferences about whether the intervention worked, how it may have worked, and which factors contributed to its success or failure. One approach for making explicit the relationship between various interacting elements of an intervention is through the development of a logic model.<sup>28</sup> Logic models are typically a graphical representation of how an intervention is supposed to work, illustrating the various inputs, activities, outputs, and expected outcomes. Such an approach provides a clear framework for monitoring and evaluating different aspects of study implementation.<sup>29</sup> In this study, we present the development of a logic model for an intervention that was explicitly developed on the basis of particular experiences and needs of the population with HD.

In therapist-led interventions, another aspect to consider is that of fidelity of intervention delivery (ie, the extent to which the intervention is delivered as intended).<sup>30</sup> Reporting of treatment fidelity is fairly commonplace in psychotherapy and counseling interventions, and specific tools have been developed for its measurement.<sup>31,32</sup> Yet, researchers have identified a failure to monitor, evaluate, and promote treatment fidelity within physical therapy trials. Hildebrand and colleagues, for instance, argue that "in occupational therapy (OT) and physical therapy (PT) outcomes research, treatment fidelity methods have not been utilized, which in our view is a serious gap that impedes novel treatment development and testing in these rehabilitation fields."<sup>33</sup> In those studies where fidelity has been measured, results have often indicated variable delivery of intervention techniques. In this study, we developed a fidelity monitoring system that included review of self-report checklists, as well as review and rating of transcribed audiotapes

from actual sessions. This rating scale enabled independent raters to determine the extent to which the intervention was being delivered as intended. The results from the independent fidelity ratings suggest that the intervention was being delivered as intended; however, coaches tended to rate themselves higher on average than the independent rater. As the coach's ratings were completed at the end of the overall study, this may have been a reflection of their increase in confidence and competence as the study progressed. Review of these audiotaped sessions also enabled the intervention coordinator to provide feedback to the coaches to make modifications to ongoing sessions.

A final important aspect that is included in this report is related to understanding the costs involved in the development and delivery of the interventions. Clearly, a full-scale health economics evaluation is imperative for phase III trials; however, we argue that preliminary costs need to be documented at an early stage in intervention development. Indeed, feasibility of an intervention should extend not only to adherence and acceptability but also to costs and training and support requirements. In our intervention, we have purposely allowed coaching staff with differing levels of skills and expertise, and some staff therefore required greater remote support in terms of training and delivery of the intervention. To further inform future implementation, we will conduct sensitivity analyses regarding staff costs, including testing the effect of using staff at a higher/lower grade to conduct the training and delivery of the intervention. The outcome of this work will be reported with the main study results. It is only by recording and considering these factors at an early stage that we can make suggestions as to the best configuration for implementation in the future.

## CONCLUSIONS

In order for a physical activity intervention to have the potential for effective translation and implementation into clinical practice, detailed information about the theoretical underpinnings, fidelity monitoring, and cost of development must be provided. This approach is novel and not yet routinely utilized in physical therapy trials. We argue here that in order for physical therapy research to take the critical steps forward in translating to the clinic, these principles must be embedded in future clinical trial designs. In this report, we have demonstrated how this can be achieved within a physical activity trial for individuals with a neurodegenerative disease. However, it is only once full-scale evaluation of the trial is complete can we then consider the potential effects of the components of the intervention, training support, and fidelity on the effectiveness of the intervention.

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where they shared their experiences of physical activity and how they could be best supported to be active with Huntington disease. We also thank Dr Fiona Jones of Bridges ([http://www.bridges-stroke.org.uk/management\\_group.php](http://www.bridges-stroke.org.uk/management_group.php)) who kindly worked with us to develop ideas "based on Bridges" that could be relevant to the HD population. We are also extremely grateful to our colleagues, from the EHDN Physiotherapy working group, Jessie van der Bent and Karin Bunnig at the Huntington-centre TOPAZ Overduin, for sharing their ideas and successes of conducting regular walking programs and other activities for people with HD.

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*Call for papers for a JNPT Special Issue on the*  
**MANAGEMENT OF COMPLEX NEUROLOGIC DISORDERS**  
**in NEUROLOGIC PHYSICAL THERAPY**

It is the patient with a rare or complex disorder that often prompts a physical therapist to turn to the literature for guidance. Regrettably, the design of larger clinical trials often excludes those with rare disorders or complex presentation. For these reasons, the investigation of these clinical populations and dissemination of the findings represents a valuable contribution to the literature, serving as a reference point for the physical therapist. This special issue recognizes the important role of evidence in the management of complex or rare neurologic disorders.

We are seeking manuscripts on topics related to complex and rare neurological conditions relevant to neurologic physical therapy. Well designed studies utilizing single subject design, small sample sizes, and case reports are welcomed.

Please note: this is a competitive call for papers. JNPT receives more submissions than can be published and it will likely not be possible to publish all manuscripts submitted in response to this call. Prospective authors are encouraged to contact the Special Issue Editors: Teresa Jacobson Kimberley (tjk@umn.edu) or Michael Schubert (mschube1@jhmi.edu) regarding their submission. First drafts will be due on June 1, 2016 with electronic publication (ePub)/MedLine indexing following acceptance, and a target JNPT print publication date of January, 2017.

Appendix 8: Example of a flyer sent out to Huntington's disease Association Specialist HD advisors to circulate



**CARDIFF UNIVERSITY**  
**PRIFYSGOL CAERDYD**

## Supporting exercise in Huntington's Disease

We are looking for up to 15 people with Huntington's Disease (HD) and/or their family members to participate in a group discussion about exercise experiences in HD. We want to know what information, training and support is needed to help people with HD to start and maintain regular exercise.

The workshop will be held on **25th April 2013 from 7pm** at the Witney Community Hospital. We will provide refreshments and we can reimburse your reasonable travel expenses.

If you would like more information, please contact Mike Cumings (Local HDA branch manager)  
Telephone: 077478413  
Or Helen Dawes (Oxford Brookes University)  
Telephone: 0186 548 3293  
Or Email:  
[hdawes@brookes.ac.uk](mailto:hdawes@brookes.ac.uk)

To confirm your attendance, please provide your name and contact details to  
Mike Cuming (HDA Local Branch Manager)  
Or Helen Dawes (Oxford Brookes University)

Please note that places will be allocated on a first come first serve basis.



South East Wales Trials Unit  
Uned Ymchwili De-ddwyrain Cymru



NISCHR  
CRC



National Institute for  
Health Research

## Appendix 9: Participant Information Sheet

School of Healthcare Studies  
Department of Physiotherapy



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Prifysgol Caerdydd  
Tŷ Dewi Sant  
Mynydd Bychan  
Caerdydd CF14 4XN

### Workshop Information Sheet

#### Title: Exercise support strategies for people with Huntington's disease

You are invited to take part in a workshop that will assist in the development of resource to support exercise for people with Huntington's disease (HD).

Before you make a decision as to whether you will participate in the workshop, it is important that you understand what it will involve. Please take the time to read the following information carefully and discuss it with others if you wish.

### Part One

#### What is the purpose of the workshop?

The purpose of the workshop is to discuss and consider how best to support people with HD to exercise in their own homes.

#### Why have I been invited to participate?

You are being asked to participate as we wish to gather the views of all relevant stakeholders. This includes people with HD, HD family members, health professionals and carers who work with people with HD and regional care advisers.

#### Do I have to take part?

There is no obligation to take part in the workshops. If you do decide to take part, you are free to withdraw at any time and without reason.

#### What will happen to me if I take part?

We will send you details (dates, time and location) of the workshops in your area. If you decide to take part, you will be asked to sign a written consent form. Any personal information will be kept confidential. All the workshops will be audio

**When completed, 1 for participant, 1 for workshop records**

Version 1; 23/01/2013

recorded to allow for transcription at a later date. The taped conversations will be destroyed on completion of transcription of the data. Written notes will be made during the workshops by the workshop co-ordinators. You may claim travel expenses incurred to attend the workshops.

**What are the possible disadvantages and risks of taking part?**

We do not expect that your participation in the workshops would expose you to any undue risk. You will be given the opportunity to discuss any concerns with the workshop co-ordinators in private and without use of audio-recording. You will be informed when any audio recording is taking place.

**What are the possible benefits of taking part?**

We do not expect any specific benefits for the individual taking part. However the information we get from this series of consultations may help us to better support people with HD to exercise in their own homes.

**What happens after the workshops have been completed?**

After you have participated in the workshops you are free to contact the co-ordinators with any question or queries you may have. If you are interested, we would be happy to send you a report of the outcomes of the workshops.

## **Part 2**

**What will happen if I don't want to attend the workshops?**

Your participation in the workshop is voluntary.

**What if there is a problem?**

If you are harmed by taking part in the workshop, there are no special compensation arrangements. If you are harmed due to someone's negligence, then you may have grounds for a legal action but you may have to pay for it. Regardless of this, if you wish to complain, or have any concerns about any aspect of the way you have been approached or treated during the course of this study, the normal Cardiff University complaints mechanisms would be available to you.

**Will my taking part in the study remain confidential?**

We would like to reassure you that your personal details would be kept strictly confidential. No one, except the named investigators, would have access to these details and no identifying details would appear in our published reports.

**What will happen to the reports that are produced from the workshop results?**

The reports or summaries thereof may be presented at conferences and published in medical or scientific journals. If you would like we can inform you of where you can obtain a copy of the published results. You would not be identified in any of the reports.

When completed, 1 for participant, 1 for workshop records  
Version 1; 23/01/2013

**Who is organizing and funding the study?**

The study and follow up workshops are organised by the Department of Physiotherapy, Cardiff University, Cardiff. Funding is provided by the NHS National Institute of Health Research.

**Who has reviewed the study?**

The study has been reviewed and ethical permission provided by Cardiff University, School of Healthcare Studies Ethics committee.

**Further information and contact details:**

If you have any questions or queries please don't hesitate to contact:

**Dr Monica Busse**  
**School of Healthcare Studies, Cardiff University, Ty Dewi Sant**  
**Heath Park, CF14 4XN**  
**Tel: 02920 687559**

Email: [busseme@cardiff.ac.uk](mailto:busseme@cardiff.ac.uk)

## Appendix 10: Consent form

School of Healthcare Studies  
Department of Physiotherapy



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Fax Ffacs +44(0)29 2068 7685  
E-mail E-bost [Physiotherapy@cf.ac.uk](mailto:Physiotherapy@cf.ac.uk)  
Prifysgol Caerdydd  
Tŷ Dewi Sant  
Mynydd Bychan  
Caerdydd CF14 4XN

### WORKSHOP CONSENT FORM

#### Title: Exercise support strategies for people with Huntington's disease.

Name of workshop co-ordinator:

Please Initial Boxes:

I confirm I have read and understood the information sheet, version 1 dated 23/01/13, for the above study and have had the opportunity to consider the information to ask questions and to have had these answered.

I understand that my participation is voluntary and that I am free to withdraw at anytime without giving any reason, this will not affect my legal rights being in any way.

I understand that all information about me will be kept in a confidential way and destroyed once the study is completed.

I understand that some conversations that take place during the workshop will be audio recorded and written notes will be made by the workshop co-ordinators.

I confirm that quotations from the workshop can be used in the final workshop report and other publications. I understand that these will be used anonymously and that no individual respondent will be identified in such report.

I confirm that members of the research team may contact me after the workshop to discuss and confirm what is spoken about and recorded during the workshop.

I confirm that I agree to my photograph being taken during the workshop which may be used in medical research publications (which may include the internet) and Huntington's Disease related newsletters and that this

When completed, 1 for participant, 1 for workshop records

Version 1; 23/01/2013

therefore may be seen by general public as well as medical professionals. *I understand that once published, it will not be possible to completely withdraw this consent.* [OPTIONAL]

I agree to take part in this workshop.

Name of subject.....

Signature ..... Date .....

Name of Witness (Workshop co-ordinator) .....

Signature ..... Date .....

When completed, 1 for participant, 1 for workshop records  
Version 1; 23/01/2013

## Appendix 11: Research ethics committee approval letter

School of Healthcare Studies  
Dean and Head of School Professor Patricia Price BA(Hons) PhD CHPsychol AFBPsS FHEA  
*Ysgol Astudiaethau Gofal Iechyd*  
*Ddeon a Pennaeth Yr Ysgol Yr Athro Patricia Price BA(Hons) PhD CHPsychol AFBPsS FHEA*



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28 February 2013

Dear Monica

### **Exercise Support Strategies for People with Huntington's Disease**

At its meeting of 26 February 2013 the School's Research Ethics Committee considered your revised research proposal. The decision of the Committee is:

#### **Pass - Proceed with Research**

Please note that if there are any major amendments to the project you will be required to submit a revised proposal form. You are advised to contact me if this situation arises. In addition, in line with the University requirements, the project will be monitored on an annual basis by the Committee and an annual monitoring form will be despatched to you in approximately 11 months time. If the project is completed before this time you should contact me to obtain a form for completion.

Please do not hesitate to contact me if you have any questions.

Yours sincerely

A handwritten signature in blue ink.

Mrs Liz Harmer – Griebel

## Appendix 12: Detailed development of the focus group schedule

The schedule began with an opening question ‘can you tell us who you are and one thing about yourself?’ going around the group one by one to introduce participants to the rest of the group. The next question was an introductory question to set the scene to the context of PA but still keeping it broad, participants were asked “What is exercise?” This was designed to establish what people conceived PA to be. Once the introductory question had set the scene for the discussion around PA and exercise, the next question was designed to contextualise and link the subject of PA to the participants. The second question linked more directly to the key questions which it led onto. The question was: “Think back to an experience of doing exercise, how would you describe it?” Krueger and Casey (2009) suggest that this question should link the subject of interest (PA in this case) to the participants. By asking them to think of their own experiences and share them it encouraged them to start thinking actively about the topic of PA from their perspective. The four questions that followed were the key questions of the FGs that were developed using the concepts identified from the literature review. These questions drove the exploration of aspects of living with HD in relation to PA.

The first key question was: *“What is it that makes you exercise?”*

The previous literature exploring physical experiences in HD pertains heavily to the concept of motivation and ‘facilitators’ to PA (Zinzi et al. 2007; Quinn et al. 2010; Khalil et al. 2012). It is seen consistently as a generic theme that could be further explored across the stages. In case participants struggled to answer and initiate or continue discussion prompts were developed that could be introduced to the group. The prompt for the first key question was: “Other people with HD who have taken part in exercise found that these things helped them to exercise: support, perceived benefits, environment and motivation. The prompts were only used if really needed which in most cases were not. The prompts were facilitators that participants of the HD exercise studies reported in helping them to adhere to the specific exercise intervention of the studies (Zinzi et al. 2007; Debono et al. 2012; Khalil et al. 2012).

Second key question: *“Think back to before you knew about the HD. How has the way you approach / think about / feel about exercise changed since then?”* This question linked into the progressive nature of HD to elicit how different aspects related to PA in HD might have changed. This pertained to the literature discussed in chapter 2 that elicited changes in abilities, and changing and negotiating different functional roles within the family, and effecting leisure and social activities, which may be of relevance to PA (Decruyenaere et al. 2003; Aubeeluck et al. 2012; Carlozzi & Tulsky 2013; Maxted et al. 2014). Also, specifying

that they should think about before HD manifested through to the present alluded to the fact that in the literature changes are seen even at prodromal stage and may elicit experiences or perceptions of how over time PA has been affected. Rather than say 'diagnosed with' using the term 'knew about' was more inclusive if people attended the FGs and were gene positive but not yet formally diagnosed with the clinical symptoms. The prompt for this question was: "Other people with HD found that tiredness, falls, social stigma of visible symptoms affected how they felt about exercising. How do you relate to this/ what do you think of this?"

The third key question was: "*Compared to other things in your daily life, where does exercise fit in?*" The third key question was developed with the salient concept of barriers that emerged from the previous literature (Zinzi et al. 2009; Elsworth et al. 2009; Quinn et al. 2010; Debono et al. 2012; Khalil et al. 2012; Frich et al. 2014). In addition more general exploration of how or whether PA was prioritised in HD and the reasons. Specific barriers identified in the previous literature were used to prompt participants if they needed it (Zinzi et al. 2009; Debono et al. 2012; Khalil et al. 2012). The prompt for this question was: "Other people found that the following limited their exercise: Holidays, healthcare appointments, caring for other family members, Domestic chores, tiredness, falls, travel difficulty, the weather, social stigma. What are your experiences?"

The fourth key question was: "*Even if it wasn't easy to do, what would help you to adapt to a new routine focussing on exercise?*"

In the literature review, the concept of 'coping' and 'strategies' was a salient feature in the literature focussing on experiences of HD. It was also explicit in the literature related to neurodegenerative studies of PA experiences but was something that had not been explored in the context of HD and PA (DI. Helder, AA. Kaptein, GMJ. Van Kempen, J. Weinman, HC. Van Houwelingen 2002; Jones et al. 2008; Dlugonski et al. 2012; Maxted et al. 2014; Skår et al. 2014). This question was developed in consideration of the fact that this may be an interesting and helpful avenue to explore especially in understanding how people may adapt PA over time across the stages. The prompt was "What strategies do you think would be helpful?" If needed further prompts were "Other people have suggested these things helped them: Somebody knowledgeable of HD, support (family members, a regular exercise group, a tailored programme) a diary to record your activity".

The FG schedule ended with the question "Is this an adequate summary?" This question followed a verbal summary given to the group by the facilitator for use upon saturation of discussion and to bring the FG to a close. Reconvening a FG at a subsequent point in time

is often impractical, and even when it can be done, the group dynamics will not be the same. Therefore, most member checking (Denzin & Lincoln 1994) needs to be done in real time while each FG is conducted. The last question facilitated member checking at the end of the FG session.

Participants of the pilot group were people with HD and caregivers that had responded to the HDA regional care advisor with the purpose of testing whether the questions flowed. (Further demographic details of the FG is given in the results chapter.) In addition, it was good practice to check that the questions were as simple and conversational as possible. This was to make sure the participants could understand them especially in consideration of possible cognitive impairments of the participants (Krueger & Casey 2009). As a result of conducting the pilot FG, the introductory question of the schedule was altered. In version one of the schedule, the introductory open question used was “What comes to mind when you hear the word ‘exercise’?” The aim of the introductory question was to establish the perspectives and PA backgrounds of the participants, and generate some initial discussion. The phrasing of this question did not facilitate that. The introductory question was changed to “What is exercise?” for subsequent FGs as it focusses on the actual activity, what people perceive to be exercise and gave an insight into participants’ views of exercise at the outset i.e. what was deemed to be exercise in their life worlds.

## Appendix 13: Focus group schedule

Opening question: Ask each person to speak in turn, say their name and whether they exercise now or have ever been involved in any kind of exercise or sport (ice breaker and to code the participants).

Once participants have introduced themselves move onto the introductory question:  
**What is 'exercise'?**

Transition Question (to contextualise/ link the subject of exercise to the participants):

**Think back to an experience of doing exercise, how would you describe it?**

Key Questions (spend most time on these):

**1. What is it that makes you exercise?**

*Prompt: Other people with HD who had taken part in an exercise programme found that these things helped them to exercise: Support, perceived benefit, environment, motivation. Introduce and discuss one at a time and use quote to support.*

*Don't use prompts straight away when asking the questions, unless you really need to but introduce them once the group has had some discussion.*

*(For example: A number of people felt that support was important to help them to do exercise and one person said this: "having someone there (at the gym) who was helping you and being completely on your side"...what are your thoughts about that?)*

**Perceived benefit:** "it's a good mood booster"; "feel more relaxed...more confident"... "Feeling fitter"... "the more I could do, the more I felt like everyone else..."; "Feel I'm walking more strongly".

**Motivation** "determined to maintain physical abilities for as long as possible"; "Coming out (after exercising) I was full of beans"

**Support and knowledge of HD:** "won't go out on her own ...I go out with her"; "(the instructor) pushed me in the best possible way"; "...the instructor at the gym itself would have to know about you..."

**Environment, social interaction:** "You have a laugh with the people there you know, I enjoyed that."

**2. Think back to before you knew about the HD. How has the way you approach / think about / feel about exercise changed since then?**

*Prompt: Other people with HD who exercised found that falls, tiredness and social stigma of having visible symptoms affected how they felt about exercising. How do you relate to this? Examples of quotes include: "falls ...put you off", "I felt a little bit exposed ... a public gym... so a bit uncomfortable."*

**3. Compared to other things in your daily life, where does exercise fit in?**

*Prompt: Other things that people found limited their exercise included the following: Holidays, Healthcare appointments, Caring for other family members, Domestic chores, tiredness, falls, travel difficulty, the weather, social stigma. What are your experiences?*

**4. Even if it wasn't easy to do, what would help you to adapt to a new routine focussing on exercise?**

*Prompt: What strategies do you think would be helpful?*

*Other people have suggested these things helped them: Somebody knowledgeable of HD, support (family members, a regular exercise group, a tailored programme) a diary to record your exercise, can you add to that? How do you agree or disagree with those?*

Give a summary of the discussion after question 4 and ask "**Is this an adequate summary?**"

Appendix 14: Demographic questionnaire for focus group participants with Huntington's disease

Please fill in some information about your self. Tick only one option.

Name: \_\_\_\_\_

1. Are you:       Male       Female

2. How old are you? (in years) \_\_\_\_\_

3. What is your highest level of education? Tick only one option.


CSE/GCE/GCSE school leaving certificate

NVQ qualification

'A' Level

University degree

Other (please specify): \_\_\_\_\_

4. Do you smoke?

Yes       No

5. How old were you when you noted the symptoms of Huntington's Disease for the first time? (in years) \_\_\_\_\_

6. How old were you when your HD was diagnosed? (in years) \_\_\_\_\_

7. Do you live? (please tick)

- By yourself
- With family
- In a residential home
- Other: \_\_\_\_\_

8. Who provides your main care?

Family member

Outside support (care package in place)

9. Do you have support to engage in social activities? (Either family member or support worker)

Yes

No

10. How much difficulty do you have with walking? **Tick only one option.**

I have no problems walking

I have some problems with walking

I have moderate problems with walking

It is very difficult for me to walk

I am unable to walk

11. Did you exercise regularly before you were diagnosed with HD?

Yes

No

12. Do you currently participate in regular exercise?

By exercise we mean specific moderate intensity exercises such as aerobic exercise (stationary cycling, treadmill), strengthening exercises, or other activities such as swimming, brisk walking, tennis, golf.

Yes

No

a. If yes to exercise: How many times a week, do you usually do 30 minutes of at least moderate physical activity or walking that increases your heart rate or makes you breathe harder than normal? **(Tick only one option)**

A few times a month

1x/week

2-3x/week

More than 3x/ week

b. Please describe the type of exercise you do (For example: weight training, yoga, walking, treadmill, golf, cycling etc.)

---

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Appendix 15: Demographic questionnaire for focus group participants (caregivers)

Please fill in some information about your self. Tick only one option.

Name: \_\_\_\_\_

1. Are you:

Male       Female

2. How old are you? (in years) \_\_\_\_\_

3. Do you smoke?

Yes       No

4. What is your highest level of education? Tick only one option.

- |                          |   |
|--------------------------|---|
| <input type="checkbox"/> | CSE/GCE/GCSE school leaving certificate |
| <input type="checkbox"/> | NVQ qualification                       |
| <input type="checkbox"/> | 'A' Level                               |
| <input type="checkbox"/> | University degree                       |
| <input type="checkbox"/> | Other (please specify):                 |

5. Do you live with a person affected by Huntington's Disease?

Yes       No

6. Are you the main carer for the person with HD?

Yes       No

7. What is your relationship with the person with HD? Please tick only one option.

- |                          |              |
|--------------------------|--------------|
| <input type="checkbox"/> | Husband/Wife |
| <input type="checkbox"/> | Child        |
| <input type="checkbox"/> | Sibling      |
| <input type="checkbox"/> | Parent       |
| <input type="checkbox"/> | Friend       |

8. How long have you been caring for an HD affected family member?

\_\_\_\_\_ years

9. Does the person that you care for have difficulty with walking?

They have no problems walking

They have some problems with walking

They have moderate problems with walking

It is very difficult for them to walk

They are unable to walk

10. Do you currently participate in regular exercise?

By exercise we mean specific moderate intensity exercises such as aerobic exercise (stationary cycling, treadmill), strengthening exercises , or other activities such as swimming, brisk walking, tennis, golf .

Yes

No

10. a. If yes to exercise: How many times a week, do you usually do 30 minutes of at least moderate physical activity or walking that increases your heart rate or makes you breathe harder than normal? (Tick only one option)

A few times a month

1x/week

2-3x/week

More than 3x/ week

B. Please describe the type of exercise you do (For example: weight training, yoga, walking, treadmill, cycling etc.).

---

---

11.a Do you currently provide support for your family member to exercise?

Yes

No

b. If you answered yes to Question 11.a. what kind of support? (E.g. Transportation, going on walks, exercising together, encouragement/reminders) \_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

## Appendix 16: Focus Group details

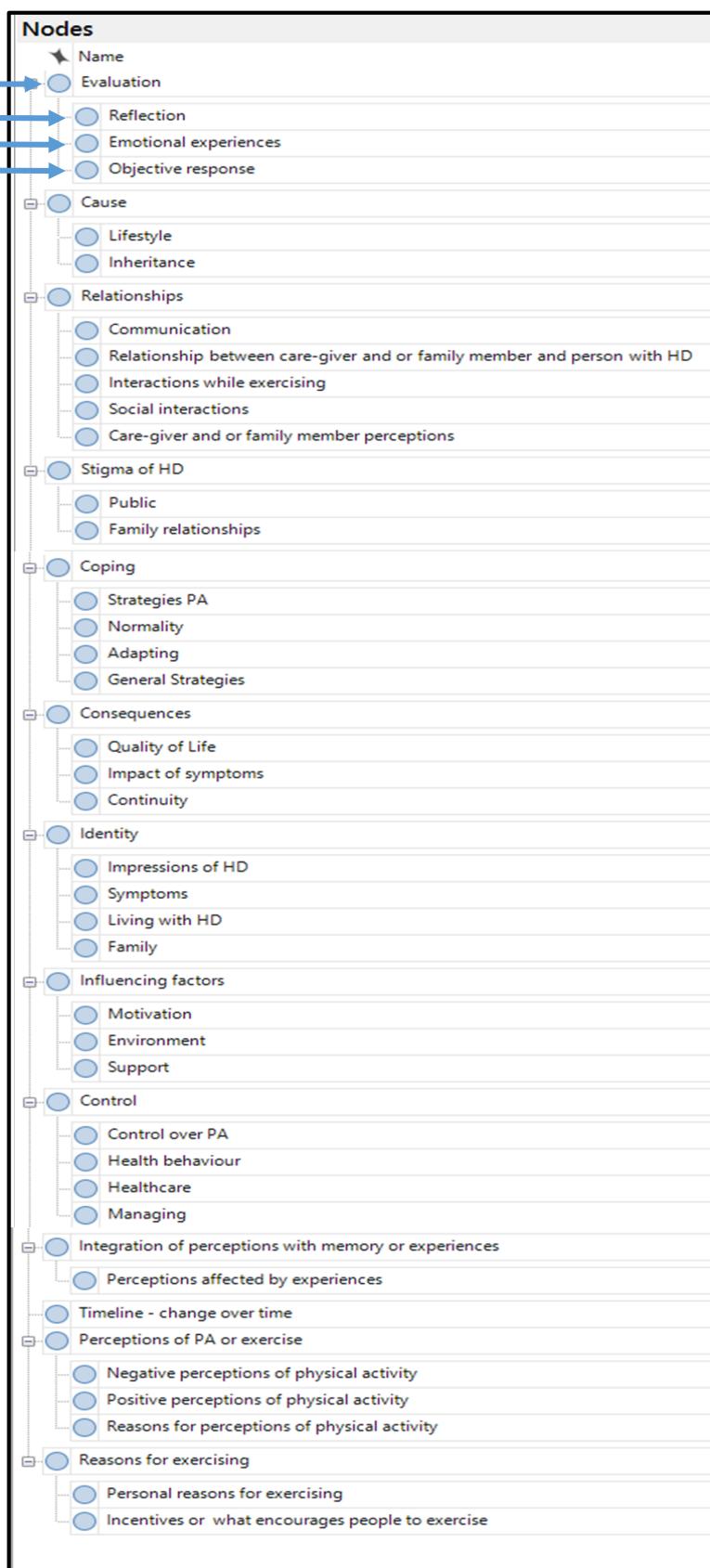
Focus Group	Site	Participants	Description
1	Suburb; affluent area of mid-England.	<b>4 people with HD:</b> 1 female (mid stage); 3 males (2 mid-late stage, 1 early stage)  <b>7 caregivers/family members:</b> 5 females; 2 males  <b>2 specialist HD advisors (1 male)</b>	A range of people with HD and family members who had previously or currently cared for people with HD participated in the FGs. There were people with early and mid-late stage HD present. One person with HD cycled regularly. Two of the family members had cared for their child who had JHD. One spouse cared for his wife and supported her to be physically active through encouragement, providing transport and engaging in research involving physical activity (PA) interventions.
2	City in the South Wales area	<b>2 people with HD:</b> 1 male early stage; 1 female mid-late stage  <b>3 caregiver/family members</b> 2 females (sister, spouse); 1 male (spouse)  <b>1 specialist HD advisor</b>	One of the caregivers had cared for her spouse for many years and he had recently moved into a nursing home because of the progression of HD. Another caregiver had been supporting her sibling for 12 years. The caregiver reported in the questionnaire that her sibling had problems with walking and did not exercise but tried to keep active. The third caregiver was the main caregiver for his spouse with mid-late stage HD who still lived at home. This person's spouse was present at the FG. She was limited functionally by poor mobility (wheel-chair to go outdoors), diagnosed 13 years ago at age 49, does not go out by themselves anymore, has outside support to do social activities, but was not exercising at the time of the FG. The couple reported that the person with HD used to exercise regularly before HD symptoms prevented them from exercising.  The other person with HD was a male at early stage and had received a diagnosis within the last 6 months at the age of 75. He reported that he had moderate problems with walking, tries to keep motivated but experiences apathy and enjoys gardening. A specialist Huntington's disease advisor (SHDA) of the Huntington's disease Association also participated in the group discussions.
3	City in the South Wales area	<b>2 people with HD:</b> 2 female's mid-late stage.  <b>3 caregivers:</b> 3 females (employed)	Three agency caregivers each had a year's experience of working with someone with HD. Two of the caregivers worked for the same person. The three caregivers were very motivated to encourage PA for the people they cared for and all three exercised regularly. The people that they worked with attended. These were two females with mid-late stage HD diagnosed 17 years ago and 10 years ago. One reported difficulty with walking and the other reported some problems with walking. One was working with the caregiver to do some PA but didn't regularly exercise before being diagnosed with HD other than skiing. The other person with HD used to exercise regularly but at the time of the FG was not participating in physical activities.
4	Urban city, northern England	<b>2 people with HD:</b> 1 female; 1 male; both early stage  <b>1 caregiver/family member:</b> 1 male (spouse)  <b>1 specialist HD advisor</b>	The Specialist HD Advisor from the Huntington's Disease Association was the advisor for the area in which the FG was conducted and exercised regularly. Of the 2 people with HD one was supported to exercise (pilates classes) by her husband (also a participant in the FG) and reported some problems with walking. The spouse had been providing some care for the last 3 years following diagnosis 4 years ago. The other person was diagnosed within the last year and had not exercised prior to diagnosis but was trying to get involved with a local Nordic walking group and went for long walks daily. He reported moderate problems with walking and lived alone. He received some support in the form of reminders to go to appointments and some daily activities from family.

5	Urban city, northern England	<b>4 people with HD:</b> 1 female early stage, behavioural symptoms; 3 males early to late stage)  <b>2 caregivers:</b> 1 female family member (mother); 1 female healthcare assistant  <b>2 professionals:</b> 1 male physiotherapist; 1 male physiotherapy assistant	The participants with HD were residents of an assisted living facility and required support with activities of daily living due to manifestation of behavioural, cognitive and motor symptoms of HD. The physiotherapist, physiotherapy assistant and healthcare assistant were employed at the home. Participants with HD all regularly participated in regular PA. The other caregiver was the mother of one of the people with HD. All of those with HD were actively encouraged and given the support to engage in a variety of regular physical activities including aerobic training, exercise classes, stationary bike cycling, football and badminton. Two people with HD exercised prior to diagnosis, 3 regularly engage in PA supported by the staff at the home. Two of the people with HD had been diagnosed 2 years previously and 6 years previously, this information is unknown for the others. One person with HD reported no problems with walking, 2 reported some problems with walking, and 1 reported moderate problems with walking.
6	Rural location, Southern England	<b>3 people with HD:</b> 2 males (mid stage); 1 female (mid-late stage);  <b>2 caregiver/ family members:</b> 1 male (spouse); 1 female(spouse)	Of the people with HD, the female was confined to a wheel chair for outdoor mobility and was older (in her 60s). Her spouse was her main caregiver who had a history of illness but now cared for her. One of the caregivers did not participate in regular exercise but did support their spouse to exercise by providing transport to attend their exercise sessions. Although 2 of the people with HD used to be regularly active with exercise and sports there had been a transition in what they do in terms of PA due to the progression of the HD.
7	Rural, affluent area of England.	<b>4 people with HD:</b> 1 female; 3 males  <b>4 care-givers:</b> 3 females(spouses); 1 male (spouse)	All participants except for 1 caregiver (spouse) were regularly engaged in PA: walking, badminton, gym sessions, golf, and swimming. All spouses were the main caregivers and all supported their husband/wife with HD to participate in PA through providing transport, walking with them, arranging the person's diary and finances for support from the NHS. 2 participants reported some problems with walking. 2 reported moderate problems with walking. 3 out of 4 people with HD exercised prior to diagnosis. Time since diagnosis was 3 to 9 years. Age range was from 47 to 63 years old. The people with HD in this group were more affected; approximately mid-stage HD.
8	Rural, affluent area of England.	<b>4 people with HD:</b> 4 males; 1 male gene positive  <b>4 caregivers/family members</b> 4 females (spouses)	All participants with HD were still independent in their activities of daily living and had active lifestyles supported by their spouses. Three people reported no problems with walking and one reported moderate problems with walking. All participants in the focus group participated in regular weekly exercise; people with HD; using a treadmill, jogging/running, attending gym group sessions, spouses; ballet, walking, going to the gym, Zumba, boxercise classes.

## Appendix 17: The analytical framework in NVivo

Adult node  
(category)

Child nodes  
(codes)



## Appendix 18: Extract of testing of the analytical framework by one of the EHDN fellows

6 4: I worry yes I'm worried he will fall *Continuity (consequences)*

7 14: Confidence can be very fragile with Huntington's

8 7: A lot of people with HD get tunnel vision...there's bugger all on either side just what's in front of ya  
9 and he'd just GO yeh, there's nothing that's where ya going, that's where ya going, doesn't matter  
0 what's happening on either side

1 12: And and who's in the way

2 7: Cars, buses, bollards, don't get it, just.... *Identity of the caregiver*

3 6: I mean you're responsible for that person and the fact that they could fall, um with *[redacted] name*  
4 especially, if he starts to fall he just lets himself go then but like if you or I tripped you control or find  
5 something to stop yourself, where as → *Impacts of symptoms*

5 7: he just goes

7 6: he just goes down yeh

3 LQ: So would you say that you sort of, he restricts activity or you're worried about him so you kind of  
9 restrict his activity because of the fear of falling, do people feel like that's the case?

1 6: Well at the moment yeh until he gets his wheelchair I wouldn't really entertain uh taking him into the  
town, I mean we go once in a blue moon now to get him down to the, the hairdressers cos he likes  
having one particular hairdresser and thank goodness the car park is at the top of where it's a  
pedestrianised area, so we don't have to worry about that so just sort of walks along n we go then to  
the hairdresser *adapting; dynamic of caregiver HD relationship*

MB: So he can walk with your help but it's the environment and the difficult negotiating

7 1 Could change over time

6 He could, he's his walking isn't that good at the moment we've got to be there not only because of  
where he thinks he wants to go, the fact he could just lose his balance *Impact of Symptoms*

MB: And then it becomes very difficult to go out because there's all these extra challenges *Environment?*

3 Sorry I think with the falling as well when [redacted] fallen it's it's trying to get back up as well  
(inaudible) managed to get back..... if I can't catch her in time then trying to get back up is an issue as  
well. → *caregivers identity & sense of burden.*

Appendix 19: Initial codes identified from open coding

Initial codes			
Communication	Outside support to exercise or do PA	Other people's attitudes to person with HD or disability/perceptions of disability	Negative perceptions of exercise
What would help them exercise - reasons for exercising	Hobbies	Understanding exercise what to do and the benefits	Carer perception of how person with HD is doing in terms of their exercise
HD community	People's goals	Challenges of dealing with other people's lack of understanding of HD	Carer's perceptions - burden or difficulties as a carer
Routine	Health of the carers who care for the people with HD	Stigma or difficulties of HD in the family	Relationship with people supervising exercise
Strategies	interaction with others while exercising	Relationship between carer and person with HD	What people with HD would like when exercising
Motivation	Supervision to ensure exercising correctly taking breaks etc	Relationship with family	Negative social interactions
External motivation including other	Worry about children inheriting HD	Exercise is about individual preference	Other people's attitudes to person with HD or disability
Terminology of exercise or PA	Need exercise specific to people with HD	Exercise or PA people enjoy doing	Research or proof of exercise working in HD
Goals or targets	What carers feel they need to be able to support the person with HD	Experiences	Language with negative connotations of exercise
Achievement	What would help them exercise - reasons for exercising	Carer's worries	Carers understanding of the individual how they function their limitations
Positive language and reinforcement from family or carer	Understanding or knowledge of other people is helpful	Social interaction	Hobbies
Ways carers or family help	Motivation of carers to help	Awareness of HD	Support looking to health and care professionals for answers
Carers perceptions limitations caused by HD	Perceived positives of exercising or PA	Co-morbidities	

Appendix 20: Example of an annotation linked to data whilst the researcher was indexing using the analytical framework

4: But I find XXX needs a lot of, umm, (pause) *positive praise for every achievement that she does because you fear that you're doing it wrong.*

5: Definitely.

4: In in everything and I don't know if that is part of the illness.

5: Yeah.

1: Do you agree with that? That it's important for you as well?

5: Defi-mmm.

4: XXX questions herself, her ability to do things.

1: Yah.

8: So literally, so if I can just really take it back to absolute basics now, so literally if you are

Annotations	
Item	Content
16	Issues of self-confidence - related to regaining identity? Reassurance she is capable. Support - verbal encouragement, positive reinforcement

## Appendix 21: Memo: 'Identity'

FG1: Across the 'Identity' codes identity as a concept does not appear to be an issue with regards to physical activity that has confronted or been considered a major issue for the members of FG1. Within the code 'living with HD' the only coded transcript is "definitely the fear of falling" (is considered a barrier to PA) but is perhaps related more to symptoms and consequences of the disease than 'identity'.

FG3 – there is one participant in particular who talks about the HD in relation to his worry about children and grandchildren inheriting it and the dependence he may have on them and how that will or does affect them. In talking about one of their sons the participant says "gets uptight for a little bit every now and then...I can see me there". The HD he says doesn't bother him so much because of his age. One thing he does say is running he's "got no chance".

Identity has not appeared as a thread for these 2 focus groups. There were more care-givers would this have an effect?

### Identity - family

It is interesting that in relation to talking about roles within the family, across 3 of the focus groups, women talked about their roles as wife and mother or their care-giver or husband did. In the context of one woman it was her care-giver who said that she constantly reminded the woman with HD that she was a wife and a mother and needed to retain these roles and maintaining her functional roles such as shopping, collecting prescriptions, baking with assistance which were all part of her physical activity plan following input from a physiotherapist. Maintaining or taking back some control over her roles within the family seemed to be a strong motivator for this woman in particular.

## FG2

**"4: I kept saying to her; you're a wife, you're a mother, you're a woman.** You need to envelope those things and become part of the family again because her husband and everybody was doing the shopping and everything for you. And now C does the weekly shop herself, umm, she goes out, she collects her prescriptions, *everything* that she needs that now has made her become a wife and a mother...again, which she wasn't having before, when we first started."

Perhaps is the female seen as the lynch pin within the roles of the family that their roles as wife and mother are highlighted and not so much the father/husband roles of the men? Or were there just more women with HD attended the groups?

## FG4

P3: "trying to maintain some sense of normality from doing those things is a big feature for M to go for that walk to be able to do it to be able to have done it is very important to you"

P2: "Yeh, yeh or just pop to the shops to get a bit of food to try and do some cooking for the kids later"

There seems to be a link here in their minds between maintaining the walking, being able to get to the shops independently and being able to provide for her children, does the word 'try' suggest hesitancy or acknowledgement that she does not always manage this? Aswell, 'trying to maintain a sense of normality' suggests that all is not right and perhaps the HD is affecting them as a family unit but they are trying to cope by trying to continue with their normal routine as much as possible. The partner of the other woman suggests that perhaps being active was a big part of the participant's role as a mother and wife and that she still wants to try to walk and stand up to do things although it is very difficult

## FG6

P7: "I think if you've been a wife and a mother you are on the go all the time and it must come as a big shock"

### Identity - symptoms

FG8 P3

"My funny walk is funny again" has HD and problematic Achilles tendon but "as long as I seem to have the right shoes then it helps with being able to do the exercise".

Aware that people might notice he is different or moves differently to others.

FG7 P11:

"Don't see them just as a HD sufferer. They need to be seen as an individual who has HD and HD is XYZ but this is the person's needs"

The symptoms of HD may have implications for a person wanting to do PA but actually the HD does not define them totally, there may be other considerations to take into account that have implications for PA. This is pertinent due to the typically sedentary behaviour of people with HD that has been reported and risks associated with physical inactivity for developing concomitant conditions.

Stand alone? Not really enough data and not strongly worded opinions – perhaps merges with impressions of HD or living with HD or rename 'living with HD' something more related to identity.

**EXPECTATIONS OF HD AND HOW THAT AFFECTS PA? INSTEAD OF IMPRESSIONS OF HD OR LIVING WITH HD? – another subtheme linked to identity?**

Impressions of HD – outside looking in?

Carer and patient impressions come out strongly worded here – relevant to FG5, FG7

### Carers

How to handle people with HD – FG5 P3 "sometimes you have to accept it and not push, cos that's the nature of her illness...in time she'll make that choice for herself, and when *she does no one will get on that bike!*"

They are people with HD – HD does not define them – they may have other problems but maybe because the motor symptoms are so visible and obvious that's all people see and not the person behind the symptoms FG7 – P11 – "...don't see them just as a HD sufferer" - links to the impressions another participant has in another group- someone with HD – the "HD ghost" FG8 P8: It's...others also assume very often when they hear about it, what it is. I call the Huntington's a ghost. Because every symptom you develop...everyone immediately sees the Huntington's

FG8 P5: Your head...I think when you find out you've got HD or you've got a gene or you're carrying it I think your head...you can let it go one of two ways. You can either let it drag you down which prevents you from doing exercise and you've got to get out of that negativity or you can run with it and try and do what P is doing and try to use it as a good thing and he's really letting his energy go you know...feeling...a lot of people I think mentally put obstacles in the way when it comes down to it. Because you feel useless I guess and you go sort of in to yourself and introverted you think...sort of...you think about yourself all the time and you become more at home. So you put obstacles in front of your exercise. You've got to overcome those as well when you find out that news. – **Questioning of oneself – could negatively affect PA if not motivate to do anything – feel 'useless' – don't know what to do...**

Under 'living with HD identity'

FG1 – generalising of people with HD ppl with HD can't play tennis because of symptoms – carer speaking (p6) "There's no way you could have someone standing up and take one foot off the floor like playing tennis!...cos over they go"

FG6 P4 "...she's been active for so many years and then having to rely on myself...you know, it gets to her, why sort of thing, sorry I can't help you"

HD causes a change in who they are as a person

Adapting to the changes caused by HD means people accepting they're not the same as they used to be and their role identities change –FG6 used to care for her husband now it's the other way around P4 (husband) "during my illness I say she's been there and helped me, say it's my turn now, innit hey"

Strong exercise identity of participant in FG5 P6 "exercise all my life, football I played, rugby I played, cricket I played, half marathons..." participant has continued to do as much as he can even though in wheelchair to go outside and living in assisted living facility.

Part of this person's social identity incorporates physical activity through dancing – going dancing in local club supported by staff from care home. FG5 participant 5

Another person sees going out as an incentive to do exercise – are they socially isolated in their home environment? FG2 P6 "I'm quite happy to go places rather than have people come to me I think me going somewhere is an incentive to do it"

Conclusion - identity is a code but needs expanding as a title for the sub theme / theme

Perhaps 'Changing identities' / fluid identities and PA – relates to illness representations? (overarching then identity as subtheme?) Identity = one of the illness representations of SRM- identity = changing over time in HD stages...theme might be changing representations in relation to HD, need to think and recheck what else is relevant here??

# PHYSIOTHERAPY BRIEFING

## Huntington's disease



### Huntington's Disease

Early physiotherapy intervention plays a key role in management of Huntington's Disease (HD) over the life cycle of the disease.

### What is Huntington's disease?

HD is an inherited neurodegenerative disease for which there is no cure. Children of an affected individual have a 50/50 chance of inheriting the faulty HD gene.

The mutation results in degeneration of the basal ganglia. This is a structure in the brain that has links to most other brain areas and is involved in coordination of body movement. People with HD gradually develop physical, cognitive and behavioural difficulties. Ability to maintain functional independence becomes limited by involuntary movements, altered muscle tone, impaired balance, and impaired speech and swallowing. Difficulties with memory, planning, prioritising and problem solving as well as behavioural issues pose an additional challenge for people with HD.

Appearance of symptoms varies between individuals but it generally affects individuals between the ages of 35-60 years of age<sup>1</sup>. Following formal diagnosis, symptoms progress over 17-20 years until full nursing care is required. Balance and mobility problems are a common feature<sup>2</sup>, leading to falls, decreased walking ability and sedentary lifestyles<sup>3</sup>.

### How can physiotherapy help?

People with HD require ongoing health and social care to support their changing needs<sup>4</sup>. Maintaining fitness and physical function from early on in the disease process is vital to maintain quality of life in HD and is especially important given the duration of the disease and known burden on healthcare services. Ongoing efforts to define physiotherapy interventions<sup>5</sup>, validate outcome measures<sup>6</sup> and conduct robust evaluations of targeted physiotherapy led interventions<sup>7-10</sup> have shown that people with HD can achieve fitness and measurable functional benefit from specific exercise training. Indeed community and home based exercise interventions have also been shown to be enjoyable, acceptable and beneficial for people with HD.

In addition, with referrals from physiotherapists, people with HD can access exercise support in local gyms through exercise referral schemes (for example the National Exercise Referral Scheme (NERS) in Wales)<sup>7,8,11</sup>. In the later stages of the disease multi-disciplinary rehabilitation, incorporating physical, occupational therapy, speech therapy and social activities has potential to moderate disease progression has been shown to be crucial to maintaining function<sup>12-14</sup>.



## **Physiotherapy should focus on maintaining fitness, mobility and function and includes:**

- Early access to evidence based self-management support to establish strategies for regular sustained physical activity<sup>10</sup> to minimise effects of primary and secondary impairments, including weakness, muscle imbalances, balance impairments and falls.
- Signposting to local support via the Huntington's disease Association of England and Wales (or Scottish Huntington's Association).
- Assistance with managing the altered walking patterns that often develop across the life cycle of disease including advice on carrying out activities of functional mobility, walking aids<sup>15</sup>, footwear and discussing adaptations in the home for optimal safety.
- Monitoring of respiratory function from the early stages of the disease to limit the potential for developing respiratory complications<sup>17</sup>.
- Encouraging independent walking for as long as is reasonably possible given the high risk of complications due to inactivity and sedentary behaviour.
- Advise carers, nursing home staff and other allied healthcare professionals on positioning and respiratory care in the later stages of the disease.

## **Case study:**

An intervention approach that promotes autonomy, competence and relatedness can improve self-efficacy for secondary prevention to slow or alter disease progression in HD. A recently completed multi-centre (8 sites) trial (ISRCTN 65378754) of an individualised life-style approach to enhancing physical activity in HD compared to social interactions resulted in Life Space scores (i.e. a measure that quantified the extent of movement within a person's environment) that were 16 points higher in the physical activity group than in the social group [95% CI 2,30]. Self-reported Physical Activity (IPAQ) scores were also 125% higher in the physical activity group [95% CI: 4%, 388%] and exercise self-efficacy was 1.63 points higher [95% CI 0.48, 2.78].

**"Early physiotherapy intervention in HD is essential to prolong fitness, mobility and function "**

## **Size of the problem:**

- Approximately 12 in every 100,000 people in the United Kingdom (UK)<sup>18</sup> are affected by HD.
- HD is dominantly inherited meaning that every person who inherits the gene will develop symptoms.
- Each child of a person with HD has a 50% chance of inheriting the faulty gene
- From formal diagnosis, duration of HD is approximately 17-20 years<sup>17</sup>.
- Aspiration pneumonia is thought to be the leading cause of death in HD<sup>19</sup>.



## Conclusion:

**Physiotherapy has an important role in encouraging and supporting people with HD to engage in activities to maintain their health, functional abilities and quality of life from early on in the disease process.**  
**Physiotherapists should be aware of the need to adapt their approach in tailoring to the individual needs of the person with HD and carer(s) who support them.**

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For more information see:  
[www.activehd.co.uk](http://www.activehd.co.uk) and <http://hda.org.uk/>



Appendix 23: Researcher's interpretation of theoretical development of ENGAGE-HD intervention

