
**A DELICATE EQUILIBRIUM:
LIVING WITH HUNTINGTON'S
DISEASE**

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ABSTRACT

Background: People with Huntington's disease (HD) can be affected by motor, cognitive and behavioural symptoms and it is this triad, coupled with its genetic nature, which makes the disease so complex, requiring multifaceted and multidisciplinary input. The length of the illness trajectory for HD can result in patients receiving care at home for an extended period during which the contribution from family caregivers is invaluable. However, there has been little research into patient or carer perspectives on needs or how these should be met, and how these correspond to professional viewpoints.

Objectives: To gain a holistic understanding of living with, caring for and working in the field of HD to:

- identify the care needs of those diagnosed with HD
- develop understanding of the needs of family caregivers caring for a person with HD
- improve understanding of the issues encountered by health and social care professionals in delivering care to people with HD
- gain an understanding of how care needs change over time
- develop understanding of care provision for those living in the family home and those in residential care settings.

Study design: The study involved people affected by HD across three geographical localities. The research took a collective case study approach with the person diagnosed with HD as the central component. Methods of data collection included the responsive interviewing technique alongside observation, eco-mapping, and genograms.

Fieldwork: These methods were used to gather data from the person with HD, their family carer and a nominated health or social care professional to build 15 cases involving 33 individuals and 115 interactions (68 interviews and 47 observations) over three years of study participation.

Findings: Living with HD requires continued readjustment to maintain balance between increasing disability, diminishing cognition and living well at home. This ongoing modification is undertaken by patients, their families and the health and social care professionals providing services and support. Patients and carers were challenged to balance: coping with the diagnosis, an impulse to secrecy and a duty to share that knowledge; autonomy, choice and decision making throughout the course of the illness; the transformation of homes to hospitals; and a shift in the burden of care when the patient moved to a residential care home. Examination of the services provided by the health and social care professionals in this study has shown how multidisciplinary working, a key worker approach, disease, person and service specific knowledge, alongside continuity of staffing contribute to quality care.

Conclusion: This is the first qualitative study of living with HD incorporating multiple perspectives over time. The study has focused on context and depth to create multiple detailed case studies. It explored the complexity of living with HD and the ways in which care can be provided in the community. It has highlighted the wider implications of the genetic nature of the condition by exploring families' decisions to share (or not) genetic information. The study has identified a number of daily challenges related to decision making and autonomy for both family and professional carers when changes in capacity occur slowly over time and are coupled with behavioural changes. Holistic, multidisciplinary and flexible care, together with a built professional knowledge and a key point of contact for families have been shown to be essential for those trying to balance the delicate equilibrium of living with HD.

PUBLICATIONS FROM THIS THESIS

Case study example for Huntington's disease in: Woodward S and Mestekcy A (2011) *Neuroscience Nursing – evidence-based practice*. Oxford: Wiley Blackwell p452, case example from Elizabeth and James.

Wilson, E, Pollock, K and Aubeeluck, A (2010) Gaining and maintaining consent when capacity can be an issue – a research study with people with Huntington's disease. *Clinical Ethics* Vol5(3):142-147 Cited on the NIHR website as 'Valid informed consent resources'

http://www.crncc.nihr.ac.uk/training/courses/other/vic_resources

Wilson, E (2009) 'Patients and families' in Byrne J, McNamara P, Seymour J, and McClinton, P (eds) *Palliative Care in Neurological Disease: A team approach*, Radcliffe, Oxford pp85-98, case example from Sarah and Max

Wilson, E; Elkan, R; Seymour, J and Almack, K (2008) A UK literature review of progressive long-term neurological conditions. *British Journal of Community Nursing* vol13(5):206-212

Aubeeluck A, and **Wilson E**, (2008) Huntington's disease. Part 1: essential background and management. *British Journal of Nursing* Vol17(3): 146-151

These publications can be found in Appendix A

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I wish to dedicate this thesis to all the families affected by HD who participated in this study. Without their willingness to devoting their precious time and energy to helping me understand a little of how they live with this condition this study would not have been possible. This thesis presents my interpretation of their stories and I can only hope that I have done them justice.

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GLOSSARY

Activities of daily living	Making meals, cleaning, laundry, managing finances
Aphasia/dysphasia	Loss or impairment of language skills required to process, understand and recall information
At risk	Those with a gene positive parent who do not know their own genetic status and therefore have a 50% chance of having the faulty gene.
Autosomal dominance	One gene is provided by the mother and one by the father to make up a chromosome pair. If a dominant gene such as HD is passed on by one parent it will block the other gene passed by the other parent so that in the majority of cases when the HD gene is passed on the person will develop the condition at some time in their life.
Bradykinesia	Slowed voluntary movements often resulting in an inability to adjust body position
Cachexia	A wasting condition characterised by a loss of appetite, fatigue and muscle atrophy resulting in significant weight loss
CAG	A trinucleotide on a gene. Expansion of the number of CAG repeats of this trinucleotide is what causes the faulty gene resulting in the development of HD
Chorea	Involuntary movements often associated with HD
CNS	Clinical nurse specialist. There is no standard definition but in Birchtree city this person was the single point of contact for patients, family carers and other health professionals. Her role was to co-

	<p>ordinate care for patients and liaise with team members. She visited people in their own homes as well as providing access by telephone. She could take referrals from any source and could refer on to additional services. She was involved with families throughout their experience with HD, including those at risk, a-symptomatic or gene negative.</p>
Dietician	<p>Provided dietary and food preparation advice and weight management. Seconded to the service in Birchtree one day per week. Was available at monthly clinics and would visit people in their own homes. Also available for referral at the other sites; a generalist without particular expertise or experience in HD.</p>
Dystonia	<p>Sustained muscle contractions that result in twisting, abnormal posture and uncontrolled movement</p>
HSCP	<p>Health and social care professional</p>
Non-penetrance	<p>The HD gene is be present in the parent but does not manifest in symptoms (this may be that they have not lived long enough for the symptoms to become apparent).</p>
OT	<p>Occupational Therapist. Provide ways to help people recover or maintain independent daily living.</p>
PA	<p>Personal Assistant. Provided to Vicky as a younger participant with no spouse to help her continue to engage with social activities and maintain her independence.</p>
PAT	<p>Pets as therapy. A national charity that provide therapeutic visits with specially trained cats and dogs to a variety of places such as hospices, care homes and special needs schools.</p>

PEG	Percutaneous endoscopic gastrostomy. A tube passed into the stomach through the abdominal wall to provide nutrition when oral intake is no longer possible or sufficient.
Penetrance	The proportion of people with the gene who develop the condition
People affected by HD	This refers to both the person diagnosed and any family members affected by having HD in the family.
Personal care	Washing, dressing, toileting
RCA	<p>Regional Care Advisor. The Huntington's Disease Association is a national charity which provides advisors to act as advocates and providers of information to people with HD and their families. The RCA in this study provided a 9am-5pm service and covered a large geographical area including Northants, Derbyshire, Leicestershire, Nottingham and Staffordshire. Oakfield town and Birchtree city sit within these areas. The role is to:</p> <ul style="list-style-type: none"> • provide information and advice to families • answer crisis calls and liaise with other professional service providers • promote and develop a full range of local services • identify suitable respite and residential care facilities • liaise with local branches and self-help groups • give talks and organise seminars and training days • provide speakers for training sessions • provide workshops for service providers and users such as health, Social Services, nursing homes, and residential care staff teams (The Huntington's Disease Association) <p>The RCA concentrated her time in the areas other than Birchtree as she was aware of the specialist HD team in the city.</p>

RSW	Rehabilitation Support Worker. A new post introduced to the Birchtree HD team two days per week in order to take on some of the work of the CNS. The person employed had a background as a physiotherapist assistant and in occupational therapy. The role involved carrying out initial assessments and follow-up visits focusing on these aspects of care.
SALT	Speech and language therapist. This therapist worked a part of a wider team for neurological conditions but had expertise and experience in HD and was seconded to the Birchtree HD team one day per week. Also available for referral at the other sites; a generalist without particular expertise or experience in HD.

The names of all persons and sites of study have been changed to ensure anonymity and confidentiality. Health and social care professionals are referred to only by professional role.

CHAPTER ONE: INTRODUCTION

Huntington's disease (HD) is a genetically inherited progressive neurodegenerative disorder. Historically cloaked in silence and shame (Hans and Gilmore 1968) our understanding of all aspects of HD is still growing. In 1972 Teltscher and Davies wrote a paper for the *Medical Journal of Australia* and stated;

"Denial of a disease is a way of life for many patients of families with Huntington's disease, ... there are popular diseases which enlist sympathy and help and unpopular diseases which interfere with jobs and personal relations." (1972, p310)

I suggest that HD remains one of these 'unpopular diseases' which over 40 years later is only now receiving attention in terms of policy direction, yet funding for care provision and understanding remains limited. Disparity between cases, even within the same family, means that experienced physicians often continue to learn from and with their patients. As Paulsen recognises in her acknowledgements

"Most of what I know about behaviour in Huntington's Disease (HD) was taught to me by the HD individuals and families I have worked with. They have struggled with me to find solutions to their problems." (2003, p1)

Due to the longevity and nature of the disease it is not feasible or desirable for a person with HD to be under constant supervision from any health or social care professional. The majority of their long-term care will be provided by family caregivers. Thus it is essential to improve our understanding of all aspects of living with the disease by engaging with the real experts; people with HD and their formal and informal support networks.

After studying medical social anthropology at undergraduate and post graduate levels I have developed an interest in people's understanding of health and illness. Anthropologists gain insight through detailed knowledge of complex real

world settings and participant experiences. This study stems from my involvement in previous research to explore the palliative care needs of service users with neurological conditions. Since joining Sue Ryder Care Centre for the Study of Supportive Palliative and End of Life Care as a research fellow in 2006 I have been involved in a number of projects exploring the patient experience of palliative and end of life care, including research with a specific focus on neurological conditions. In the course of undertaking research with patients in long term care, I became increasingly interested in understanding the particular needs of people with HD. The complexity of the condition and its genetic nature pose a number of challenges for patients, families and health professionals making it intrinsically interesting. HD is also under-researched, so an important area to investigate. In the current study I have drawn on my background in anthropology and research in nursing studies to develop a strong methodology for undertaking ethical and sensitive research in this challenging area of patient experience of disease and health care. Case studies were a small element of the earlier study, but offered an approach which I felt was the best way to gain an understanding of the multifaceted nature of HD. This doctoral study therefore develops the case study method to incorporate a greater number of cases and an extended period of participant follow up. It also includes community and hospital-based HD services alongside the long-term care centres.

Chapter Two of this thesis expands on this brief introduction by reviewing the current literature on HD. The first section of the chapter addresses how HD affects the brain, its subsequent implications for motor, behavioural and cognitive symptoms and the wider consequences of such a complex condition including coping with a chronic illness and its impact on autonomy and decision making. A brief overview of the stages of the condition, the background to the genetics and genetic testing, and the epidemiology of disease is then provided. Focus is then turned to current policy and guidance for neurological conditions, and where it exists, for HD. The final section presents the limited literature on service provision for HD. It addresses multidisciplinary working, specialist nursing input, the role of family caregivers, self-management and how a

palliative care approach is being increasingly drawn upon to inform practice in HD service provision.

This thesis reports on a longitudinal study of fifteen cases incorporating people with HD, their family carers and the health and social care professionals they engaged with over time. Chapter Three describes how a case study approach has promoted different methods of qualitative data collection from these different perspectives. Interviews, observations, eco-mapping, and genograms were all used to build a comprehensive picture of living affected by HD. Chapter Three focuses on the research design, the methodology of the study, the methods of data collection and the methods of analysis.

Chapter Four serves to contextualise the findings and provide a link between the individual case profiles provided in Chapter Five and the three qualitative findings chapters (Six, Seven and Eight). Chapter Four is divided into three main sections. Firstly the three sites where the study took place are described. Then data which form the demographics of the cases are presented in order to show recruitment, study duration, study participation, data collection and prior knowledge of HD in the family. The third section reflects on the methodological learning from undertaking the study, and goes on to discuss how findings have been interpreted and the potential validity of those interpretations.

Having shown how the cases have been built from the demographic data Chapter Five presents individual case profiles in order to adhere to participants' stories in the context in which they were told. The case profiles introduce summaries of each case in alphabetical order. They including those involved in the case and the types and frequency of data collected. This information is provided alongside genograms to depict genetic family histories. Samples of eco-maps are used to show the number and type of people involved in the person's care and how these changed over time. Information about living situation, level and type of care input and key information points are also provided.

Chapters Six, Seven and Eight present the qualitative data from interviews and observations. These three chapters are broadly based on the early, mid and late

stages of the disease. They draw on examples from cases considered to fit within these broad stages. A table at the beginning of each chapter shows the individual cases drawn upon to illustrate points in that chapter. As the participants were in the study for a period of up to three years their disease progressed over time and some are used as examples in more than one chapter.

A theme apparent throughout the data and across all stages was one of balance. Therefore, each findings chapter is illustrated by a diagram at the start of the chapter to show the tenuous balance people strove to maintain during the different stages of the disease. As the condition progressed an increasing number of services were put in place in order to maintain this balance to allow the person with HD to continue to live well at home. Chapter Eight shows that family carers were a key part of this fragile equilibrium. Once family carers were no longer able to cope with the increasing cognitive, behavioural and physical deficits of the disease the balance was finally tipped and the person moved to full-time care provision.

Chapter Nine provides a discussion of the issues affecting the delicate balance of living with HD. It is divided into two sections to address the key findings for patients and families, and the health and social care professionals. It is positioned in conjunction with the relevant literature and current UK policy. The first section addresses how patients and families respond to the new knowledge of a diagnosis of HD, the burdens this places on them to decide whether or not to share this information, the management of complex conditions in the home environment and how the 'home' is altered by equipment, supplies and adaptations. This section on patients and family carers also addresses the complex issues of autonomy, capacity and choice and how family carer burden is not fully lifted once a spouse moves to residential care. The second section in the discussion chapter focuses on the issues for health and social care professionals and key aspects of quality service provision. This is summarised in a model for HD and attention is paid to the key worker role, specialist knowledge, continuity of care and the potential for self-management. These arguments are then drawn together in Chapter Ten where the

contribution to new knowledge is recognised alongside areas warranting further exploration and research.

CHAPTER TWO: LITERATURE REVIEW

INTRODUCTION

This chapter describes how HD affects the brain and the complex and multifaceted symptoms that can manifest throughout the progressive phases of the condition. Some of the wider implications of the condition are discussed with a focus on the implications for families, the theory around coping with a chronic illness and the impact on capacity and autonomy. The key issues from the clinical and academic literature relating to the genetics of the disease, testing and the potential impacts of the test are then highlighted. This chapter then presents an overview of the epidemiology of neurological conditions by drawing on the Global Burden of Disease study (Murray and Lopez 1997). The final two sections of this chapter address UK policy, guidance for neurological conditions, and service provision.

Although bestowed with the name of the disease, George Huntington's 1872 paper was not the first written record of HD. In 1841 Charles Waters and in 1860 Johan Christian Lund also described a hereditary disease with chorea like movements, evident in adult life and slowly progressive (Siemers 2001). However, it is Huntington's paper 'On Chorea' (2003 originally 1872) that provides the most comprehensive and irrefutable description of hereditary chorea;

"... is confined to certain and fortunately a few families, and has been transmitted to them, as an heirloom from generations away back in the dim past. ... It is attended generally by all the symptoms of common chorea, only in an aggravated degree hardly ever manifesting itself until adult or middle life, and then coming on gradually but surely, increasing by degrees, and often occupying years in its development, until the hapless sufferer is a quivering wreck of his former self. ... There are three marked peculiarities in this disease: 1) Its hereditary nature. 2) A tendency to insanity and suicide. 3) Its manifesting itself as a grave disease only in adult life... I have never known a recovery or

amelioration of symptoms in this form of chorea; when it once begins it clings to the bitter end. No treatment seems to be of any avail, and indeed nowadays its end is so well known to the sufferer and his friends that medical advice is seldom sought. It seems at least to be one of the incurables” (Huntington 2003 originally 1872, p111).

HOW HD AFFECTS THE BRAIN

HD affects parts of the basal ganglia in the brain called the caudate nucleus and the putamen (Quarrell 1999). The basal ganglia is located deep within the brain and is particularly important as it organises the motor movements of the body. The caudate’s vital role is to relay messages from one part of the brain to another, regulating the information that passes through (Paulsen 2003). Its connection with the frontal lobes means that it influences motor skills, cognition and emotions and mood. HD causes the caudate to atrophy, stopping the circuits in the brain working so that messages cannot be transmitted from one area of the brain to another. When this happens people with HD become unable to control movements, thoughts and feelings. This triad of symptoms as shown in Figure 1 have been noted and recorded since early identifications of the disease.

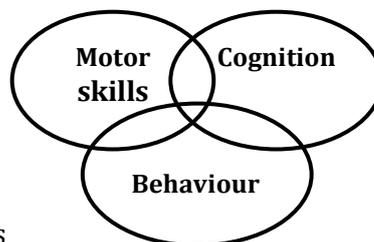


Figure 1 - Triad of symptoms

Although these symptoms are well characterised there remains uncertainty as to sequencing and progression of the symptom presentation, particularly in the early stages (Kirkwood et al. 2001). As Kirkwood et al. (2001) identify, there is conflicting evidence as to whether motor or cognitive and behaviour symptoms denote the onset of the disease. Chorea is often reported as the

presenting symptom but further interrogation can reveal mood, personality, and cognitive change such as irritability, depression and a lack of concentration often precedes movement disorders (Halpin 2011). Current research is also showing changes in the brain occurring decades before formal diagnosis of motor symptoms or recognition of cognitive changes. These changes produce cognitive changes as well as motor and psychiatric symptoms (Harrington et al. 2012). In the UK this is part of ongoing work being conducted by the PREDICT-HD team. Findings have yet to be published (Tribrizi 2012). The majority of research on HD is focused on the search for cure by better understanding the changes in the brain and the genetic aspects of the disease.

MOTOR SYMPTOMS

Chorea is the characteristic symptom of HD which features uncontrolled involuntary movements which can start out as tics, twitching or inability to sit still. Bradykinesia, slowed voluntary movements and dysytonia, slow and prolonged muscle contractions (Quarrell 1999) are also key features of motor disturbances of HD.

As laryngeal and respiratory muscles become involved in the movement disorder both speech and swallowing are affected (Simpson 2004). The rate and rhythm of speech are disturbed. This is coupled with reduced cognitive function which impairs ability to find the right word, or recall information.

COGNITIVE ASPECTS

Cognitive functions are impaired making it increasingly difficult for the person to take in information via the senses and to process, store and then recall that information (Bourne et al. 2006). This affects concentration, attention, organisational skills, spatial awareness and memory. One of the most difficult aspects for families is the inability of the affected person to recognise how their symptoms are affecting themselves and the people around them (Bourne et al. 2006). People with HD are often unable to assess the consequences of their actions and often overestimate their abilities, while

not recognising their deficits due to the disease (anosognosia) (Simpson 2004).

BEHAVIOURAL ASPECTS

Cognitive and behavioural elements are interlinked as cognitive impairments often give rise to behavioural problems. Cognitive changes impair the ability to take in information and adapt to new situations. This often results in fixed thinking, requiring adherence to rigid routines (Bourne et al. 2006). This rigidity of thinking is often expressed as performing the same activity over and over. However obsessive-compulsive disorder (OCD) as a condition in itself is not common in persons with HD (Ranen et al. 1995). Slowed thinking/responses, memory problems, and difficulties in sequencing activities can all be apparent. Visuospatial awareness is often affected, resulting in what can be perceived as aggressive behaviour as the person is no longer able to accurately judge distance so may, for example, stand too close or slam a cup on the table (Bourne et al. 2006).

Impulse control, irritability and temper outbursts can all be factors of affected behaviour and are often an outlet for frustration at the slowed thinking caused by the disease. An environment of routine and limited distraction can help manage these behaviours (Paulsen 2003). Apathy is also a key symptom of HD and can cause significant problems for the person with HD and the people around them. It involves elements of lack of energy, poor self care, failure to complete tasks, emotional blunting, lack of initiative and poor quality of work (Naarding and Janzing 2003, Thompson et al. 2002). Although sharing common features with depression, apathy is a distinct element of the disease process identified in more than two-thirds of participants in Craufurd et al.'s (2001) study.

STAGES OF HD

There are three main stages to the progression of HD: early, middle and late (Kent 2004, Ranen et al. 1995, Kirkwood et al. 2001). Early stages manifest as some initial involuntary movements or twitching, often coupled with some intellectual and personality changes such as depression, irritability, some difficulty processing information and carrying out complex tasks. Initially minor cognitive changes may only be recognisable by close family members (Williams et al. 2007) and will often be attributed to other things such as reduced memory, or an inability to learn new things may be attributed to advancing age. It is often only when motor symptoms become apparent that people seek medical advice.

During the mid stages motor abnormalities become more visible (Aubeeluck and Wilson 2008) presenting as involuntary movements and impaired voluntary movements. These affect gait, fine motor control, reaction times and often result in falls, dropping things and general clumsiness. Speech also becomes increasingly impaired as does swallowing resulting in a need for revised dietary and communication methods. During the later stages of the disease full care is required as the person becomes largely unable to speak and bedridden. Chorea, or more commonly rigidity and bradykinesia, are more pronounced, coupled with global dementia (Kirkwood et al. 2001, Ranen et al. 1995). Cachexia can also be an issue in the later stages of the disease (Kent 2004).

People with HD present with a range of complex and challenging symptoms, some of which have been highlighted in this section. Symptoms can vary from person to person as well as throughout the course of the disease, sometimes disappearing only to be replaced by another. Even the signature choreiform movements are not uniformly present, resulting in a shift towards the name Huntington's 'disease' rather than 'chorea' (Teltscher and Davies 1972). Over time the number and severity of symptoms increases, requiring more and more skilled management. As the Huntington's Disease Society of America indicate:

“There’s no typical person with HD. Each individual has complex, unique needs. Some needs can be met easily. Others will require clever or creative solutions. Still others will require ongoing trial-and-error approach.” (Huntington’s Disease Society of America 1999, p7)

It is these complexities, uncertainties and the unpredictable nature of the condition that make caring for those with HD such a challenge.

GENETICS

HD is a dominant autosomal gene, with its mutation located on chromosome 4. Humans have 23 homologous chromosome pairs, 22 of which are autosomal (non-sex) chromosomes and two distinct sex chromosomes (X and Y). Located on an autosomal chromosome HD is not sex linked and therefore affects males and females equally. Approximately 20-25,000 genes are located on the chromosome pairs. A person inherits one chromosome of the pair from each parent. A dominant gene on that chromosome will then effectively shut out the instructions of the other. For example if a child is born to a couple where the father has HD and the mother does not, one chromosome from the mother’s pair and one from the father’s pair will then make up the new pair in the child. Both the mother’s chromosomes are healthy so it does not matter which is passed on to the child. However one of the father’s chromosomes carries the gene for HD giving the child a 50% chance of inheriting the chromosome with the HD gene and a 50% chance of inheriting the normal chromosome from the father. If the normal chromosome is passed on then there is no risk of developing HD. However, as the HD gene is dominant, if the affected chromosome is passed on to the child in the majority of cases they will develop the disease providing they live long enough (Brouwer-DudokdeWit et al. 2002). Hence the penetrance (the proportion of people with the gene who develop the condition) was thought to be almost 100% (Quarrell 1999). Whether a person has the mutated gene which causes HD is dependent on the number of CAG repeats located on one

of their inherited chromosome pairs. The length of the CAG repeats affects the size of the gene and the level of mutation. Better understanding of these repeats in recent studies have shown that for those with a CAG repeat length between 36 to 39 penetrance is reduced (Walker 2007).

There is no clear age of onset for the disease. Experts usually average this to be between 35 and 55 years of age (Quarrell 1999). Due to the insidious nature of its onset it is also difficult to estimate the duration of the disease (Pakenham et al. 2004) and this is often placed at between 10 and 20 years (Houlihan 1999).

TESTING

Genetic testing for the mutation that causes the condition is a relatively new process made available in 1993 (Brouwer-DudokdeWit et al. 2002, Pakenham et al. 2004). Prior to this linkage was used to predict the likelihood of developing HD (Meiser and Dunn 2000). The genetic test can now be used to confirm a diagnosis of HD or predict the presence of HD in a-symptomatic individuals considered to be 'at risk' because of their genetic link to someone with HD.

The uptake of the predictive test since 1993 has been considerably lower than expected (Brouwer-DudokdeWit et al. 2002, Maat-Kievit et al. 2000, Meiser and Dunn 2000, Pakenham et al. 2004, Houlihan 1999). In their review of the literature on the psychological impact of testing Meiser and Dunn (2000) identify uptake of the test to be between 9%-20% globally and Maat-Kievit et al. (2000) between 2%-16%.

The test itself only takes a few days, however genetic counselling is a vital part of undertaking the predictive test and must involve more than one session over at least one month (Quarrell 1999, Maat-Kievit et al. 2000). The counselling focuses on the whole family as well as the at-risk individual. Its role is to ensure a person wishing to be tested has all the necessary information to make an informed decision. Even those known not to be at risk, for example those with a negative test result or non-genetic family

members such as a spouse, can benefit from genetic counselling as there are many issues raised by both a positive and negative test.

Whether a person has the mutated gene which causes HD is dependent on the number of CAG repeats which affect the size of the gene. There is, as shown in Figure 2, a small overlap between the normal sized gene and the HD size therefore there are four possible outcomes to the test (The American College of Medical Genetics and American Society of Human Genetics Huntington's Disease Genetic Testing Working group 1998):

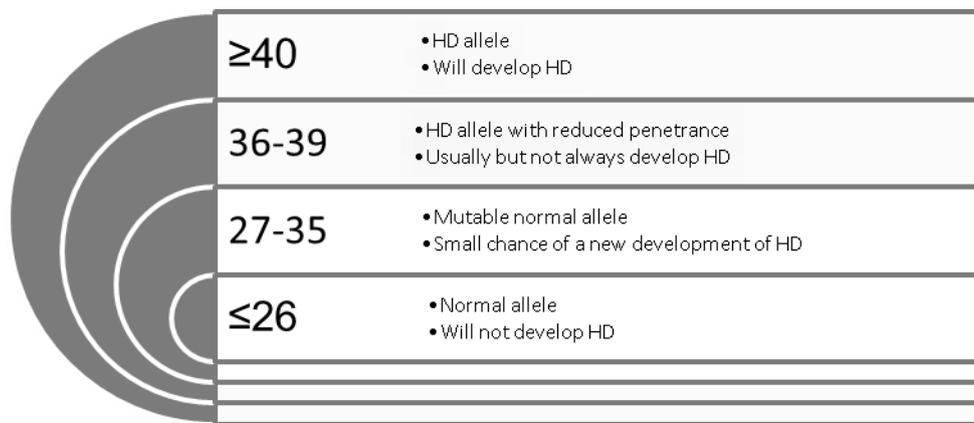


Figure 2 - CAG-repeat length categories

There is evidence that the larger the number of repeats in the CAG length the earlier the onset of the disease (Squitieri et al. 2008). However this is not yet a conclusive predictor, only directing to a potential age range for disease onset. As such it is not standard practice in the UK to inform people being tested of their CAG-repeat length.

Halpin's recent paper addressing the divide between neurology and psychiatry touches on the issues of misdiagnosis and its impact on the individual and future generation (2011). A key impact of misdiagnosis or no knowledge of HD in the family is that it can delay the correct diagnosis for those currently experiencing symptoms of HD as physicians may not know to test for it. Almquist et al. (2001) found that 44% of people newly diagnosed with HD did not know of the condition within their family and calculate 8% of

these to be new mutations (also see Ramos-Arroyo et al. 2004). There are several other reasons a person may have no knowledge of HD in their family including the early death of family members, previous misdiagnosis, non-paternity, fractured biological families and non-penetrance of the disease (Almqvist et al. 2001, Halpin 2011).

Reluctance to acknowledge the condition and secrecy within families can also prevent information being passed to future generations and result in a person being unaware that HD is in their family. Hans and Gilmore suggested two forms of denial in their paper of 1968. The first is in line with Paulsen's (2008) suggestion that denial or a lack of openness is an expression of fear of rejection or ostracism. Families hide the existence of HD in order to protect themselves socially, for employment, and for financial resources such as insurance and mortgages. The second form of denial suggested by Hans and Gilmore (1968) is one of self-protection. Children may ignore or deny the disease to suppress fears of their own risk, or a parent may feel guilty for passing on the condition. Secrecy, silence and shame still surround the disease and families may conceal its existence in order to avoid rejection or discrimination (Paulsen 2008). This has often been built on years of negative medical histories, stereotyping, and misrepresentation of HD (Wexler 2010). For example the association with witchcraft and criminality, despite being debunked, is a powerful legacy (*The Lancet* 1933, Vessie 1932) and discrimination remains rife today (Bombard et al. 2009, Bombard et al. 2007, *The Lancet Neurology* 2010). However, it is not just genetic testing that contributes to such discrimination. As Tibben (2009) suggests, years of family experiences and subsequent narratives surrounding HD also have an effect on people's perceptions of how their family has been treated over the generations.

IMPACT OF TESTING

The genetic implications of HD have a number of consequences for the person with the gene and their family members. Much debate has surrounded genetic testing for the gene since the initial markers were

identified. It is beyond the scope of this thesis to do more than summarise some of the issues discussed in the current literature.

The reasons why people choose not to be tested are complex but the psychological and emotional impact of a positive result are often cited (Meiser and Dunn 2000, Maat-Kievit et al. 2000). Other reasons included the lack of a cure or treatment for the condition and implications for health insurance and finances. For those who already have children, being tested would confirm the level of their children's risk (Meiser and Dunn 2000, Maat-Kievit et al. 2000, Mayor 2000). Codori and Brandt's (1994) participants also identified the fear of being a burden in the future, a sense of guilt for passing on the gene, increased symptom searching and a preoccupation with developing the disease as the negative consequences of receiving a high risk result (Codori and Brandt 1994). Confirming someone has not inherited the HD gene can be as emotionally distressing as a positive result, for different reasons. Survivor guilt is often an emotion experienced by family members who have tested negative when others have received a positive test (Meiser and Dunn 2000, Maat-Kievit et al. 2000, Codori and Brandt 1994). Furthermore, several areas of debate surround the legalities of ownership of genetic information (Lucassen and Clarke 2007, Hakimian 2000, Huniche 2011).

Some positive effects of testing were also identified by Codori and Brandt (1994) and Hagberg et al. (2011). Codori and Brandt (1994) surveyed high and low risk participants who had undergone linkage testing for the HD gene. Whether their result was positive or negative, respondents felt they benefited from knowing what the future held, allowing them to spend less time worrying about developing HD. Those found to be the gene carrier had benefited from being able to realise what is important in life (53%), travel more whilst they could (29%) and to make financial preparations for the future (24%). Those not at risk were able to engage in less symptom searching (71%) and tell their children they were not at risk (80%) (Codori and Brandt 1994).

WIDER IMPLICATIONS

It is not just the physical aspects of HD that make the disease so difficult to manage. Social, financial and emotional strains are also borne by families affected by the disease. In a small number of instances aggressive and compulsive behaviours can result in anti-social behaviour or criminal activity (Bourne et al. 2006, Paulsen 2003). In the pre-diagnosis stage this can lead to the involvement of mental health services and policing. To the uninformed, people with motor problems may be assumed to be drunk, associating a negative stigma with the condition (Halpin 2011, Teltscher and Davies 1972).

The financial implications can be enormous. As the condition progresses not only can the person with HD not work, but often the partner or spouse of that person must also give up work to become their full time carer (Aubeeluck and Moskowitz 2008). A dual earning family can be reduced to no income other than state support. Restricted incomes can also mean that even if families could take respite from their caring roles, a holiday is not financially viable. Impact on family carers is essential to consider as their own health and welfare is often affected by their caring role (Williams et al. 2012, Williams et al. 2009, Aubeeluck 2005, Aubeeluck and Buchanan 2006, Aubeeluck and Moskowitz 2008, Semple 1995, Tyler et al. 1983). In a Scottish self-report survey, which included carers, the research found 45% of carers were identified by the Hospital Anxiety and Depression Scale as having clinical anxiety/depression. Seventy-five percent reported perceived stress, 81% had comparably poor health and 70% had a comparably poorer lifestyle when their levels of psychological distress were compared to others in their age group (McGarva 2001). The 'home' can also be transformed into a caregiving space, encumbered by equipment and accessed regularly by strangers in the guise of health and social care professionals (Dyck et al. 2005).

COPING WITH CHRONIC ILLNESS

Chronic illness has been described as a biographical disruption (Locock et al. 2009, Williams 2000, Bury 1982). Illness itself can create initial physical

dependency but, with health care interventions, potential for recovery. Chronic illness only allows for these features in part. It is directed by the prospects of limited recovery and increasing dependence. Bury suggests that the onset of a chronic illness represents 'an assault not only on the person's physical self, but also on the person's sense of identity...' (Bury 1991, p453). This thesis does not allow for a full discussion of all the concepts around chronic illness raised in the sociological literature. However, the concept of disruption and the possibility for adaptation are potentially relevant to those affected by HD. In their study of the stories told by people with Motor Neurone Disease Brown and Addington-Hall (2008) identified four narrative types: sustaining, enduring, preserving and fracturing. The sustaining narrative was conceptualised as embracing what remained positive in life by living well for as long as possible, keeping active and remaining socially engaged with life. The enduring narrative identified the person as disempowered, viewing the situation as insurmountable. The preserving narrative focused on actively undertaking endeavours to increase survival and fight death. A feature of this narrative was the swing between hope and despair. Finally, the fracturing narrative was identified by those who talked of their loss of future as completely shattering, and being overwhelmed by concerns of loss and fear of what was to come (Brown and Addington-Hall 2008).

Stories of coping with chronic illness may depend on the type of chronic illness participants were dealing with. Robinson's (1993) paper offers a concept of 'normalisation' when managing a chronic illness such as asthma, muscular dystrophy, multiple sclerosis, back problems, or heart disease. This dominant story of living life as normal or a notion of 'normal illness' (Williams 2000) is similar to Brown and Addington-Hall's (2008) sustaining narrative and may be a useful strategy for dealing with a progressive and complex condition such as HD.

CAPACITY AND AUTONOMY

There is now increasing evidence that changes in the brain take place long before symptoms of HD become evident (Harrington et al. 2012, Tribrizi 2012).

These prodromal symptoms not only produce subtle motor and psychiatric symptoms, but cognitive changes. Changes then continue throughout the disease trajectory affecting cognition in a number of ways including understanding and decision making. The Mental Capacity Act has been in force since 2007 to support people to make their own decisions (Department of Constitutional Affairs 2007) and is based on a presumption of capacity unless proven otherwise.

The Mental Capacity Act is underpinned by five principles:

- 1) A presumption of capacity – every adult has the right to make his or her own decisions and must be assumed to have capacity to do so unless it is proved otherwise.
 - 2) Individuals being supported to make their own decisions – a person must be given all practicable help before anyone treats them as not being able to make their own decisions.
 - 3) Unwise decisions – just because an individual makes what might be seen as an unwise decision, they should not be treated as lacking capacity to make that decision.
 - 4) Best interests – an act done or decision made under the Act for or on behalf of a person who lacks capacity must be done in their best interests.
 - 5) Least restrictive option – anything done for or on behalf of a person who lacks capacity should be the least restrictive of their basic rights and freedoms.
- (Department of Constitutional Affairs 2007)

Importantly, the Mental Capacity Act allows for an appointed person to make decisions about that person's health care should that person lose capacity to do so for themselves. Some concerns have been raised about this widened legislation and burden of responsibility for those appointed to make these 'proxy' decisions (Schiff et al. 2006). Schiff et al. (2006) also cite the lack of evidence that decisions made by patients and proxies concur, the potential for conflicts of interests, and the need for a system to resolve situations where clinical staff do not feel a decision has been made in the patients' best interest as additional concerns. Ho (2008) discusses the importance of the family's role in decision making by drawing on the concept of relational autonomy. She argues

that patients are not independent units but function in the context of social and family histories (Lloyd et al. 2012, Nolan et al. 2004). Respecting the family unit in this way preserves 'an overall sense of identity, agency and selfhood' (Ho 2008, p131) through their connections with others by recognising family members as consistent and embedded among the, often interchangeable, range of health and social care professionals. Ho (2008) suggests that health and social professionals therefore need to recognise that patients and their families have 'overlapping considerations' (p131) and 'intertwined lives' (p132) which affect their priorities and the ways in which they make decisions. Patients therefore, may wish to actively engage family members in decision making and, or put the needs of their family above their own.

The Mental Capacity Act (2005) also provides for people who anticipate a loss of capacity at some future time to draw up an 'advance decision' to refuse specified medical treatment in particular future circumstances (Wilson et al. 2010b). An advance decision 'enables someone aged 18 and over, while still capable, to refuse specified medical treatment for a time in the future when they may lack the capacity to consent to or refuse that treatment' (Department of Constitutional Affairs 2007). An advance decision will only come into effect when the individual has lost capacity to give or refuse consent to treatment (Halliday 2009).

There has been little to demonstrate what the effects of the Act have actually been for health and social care professionals working within this legislation. A study by Wilson et al. (2010b) identified the need for a clearer understanding of the Mental Capacity Act, and particularly the terminology of the Act in order for staff to hold discussions with patients and families for care planning. A report from the University of East Anglia provides some interesting insight into the impact of the Act on the development of social workers' decision-making (McDonald et al. 2008). Social workers working with people with dementia were asked to reflect on the impact of the Mental Capacity Act on their casework and record keeping. Findings showed that the implementation of the Mental Capacity Act had significantly impacted the structure of decision-making for the social workers, helping them to develop greater confidence in their assessment skills. Recognition of fluctuating capacity, involving

people in their own decisions, and multidisciplinary working were highlighted, as was the potential for the social workers to take on roles as advocates and legal representatives (McDonald et al. 2008).

Previous research with staff providing care to people with progressive long-term neurological conditions suggests capacity should be assessed according to the level of decision needed to be made (Wilson et al. 2010b). For those with HD capacity is likely to diminish slowly over many years and may be valid for some decisions but not others. Identifying how and when someone no longer has capacity to make a decision poses considerable challenges for those providing care.

EPIDEMIOLOGY

The Global Burden of Disease study suggests that the burdens of neuropsychiatric disorders are under recognised (Murray and Lopez 1997) and that neurological disorders as a whole have emerged as a leading health problem worldwide (Menken et al. 2000). Based on a disability framework rather than mortality statistics alone the study was able to calculate years lived with a disability as well as years lost through premature death. In 1990 neurological conditions accounted for 28% of years lived with disability despite only accounting for 1.4% of deaths. This is continuing to rise, making neurological and psychiatric disorders the most important contributors to years lived with disability across most of the world (Menken et al. 2000). The authors recommended that neurological disorders be made a worldwide priority with services prepared to deal with increasing numbers of patients.

The scope of the term 'neurological conditions' covers hundreds of different conditions and diseases from headache to acquired brain injury. In the UK HD is categorised as a progressive long-term neurological condition by The National Service Framework (NSF) for Long-term Conditions (Department of Health 2005). In such categorisation 'progressive' long-term neurological conditions are different from those that are stable, such a post-polio syndrome or adult cerebral palsy; the result of sudden onset, such as stroke

or acquired brain injury; or those that are intermittent or relapsing, such as early Multiple Sclerosis or epilepsy (Department of Health 2005).

No accurate numbers for how many people have Huntington's disease are available, but in 2010 the Huntington's Disease Association registered 6702 symptomatic people in England and Wales (Rawlins 2010). A document by the National End of Life Care Programme on the number of deaths from neurodegenerative disease in the UK reports 236 deaths in 2008 where HD was cited in the cause of death (National End of Life Care Programme 2010). Although incidence is unknown prevalence is estimated at 13.5/100,000 population in the UK. This can be compared to 114/100,000 population for Multiple Sclerosis, 7/100,000 population for Motor Neurone Disease (The Neurological Alliance 2003) and 825/100,000 population for cases of cancer (National Statistics 2007). Despite potential underestimations of the numbers of people with HD and at risk it remains a rare disease (Rawlins 2010, Spinney 2010).

POLICY AND GUIDANCE

The National Service Framework for Long-term Conditions (Department of Health 2005) due to be implemented by 2015, is presented as a key tool for delivering the government's broader strategy to support people with long-term conditions. It provides quality requirements for all aspects of services, and is supported by the White Paper: *Our Health, Our Care, Our Say: a new direction for community services* (Department of Health 2006), which sets out a vision to provide people with good quality social care and NHS services in the communities where they live.

In the UK initial focus on neurology brought about by the introduction of the National Service Framework has been renewed by concerns that implementation of the framework has not been adequately achieved and several of the quality requirements remain unmet (Neurological Commissioning Support 2010). A scoping exercise published as early as 2008

highlighted concerns that wider policy initiatives were not taking the National Service Framework into account and that neurology remained overlooked in ongoing changes to health and social care services (Bernard et al. 2008).

A further report by the Royal College of Physicians (2011) highlighted the shortage of neurologists in the UK in terms of meeting the demand for services and in comparison to other European countries. The report also suggests that improved cooperation between neurology and psychiatry is necessary to meet the needs of those with multifaceted conditions such as HD. Again noting the failure to meet previously made suggestions concerning neurological services, the report continues to identify several recommendations specifically for long-term neurological conditions. Recommendations focus on joint planning across care providers, the appointment of clinical leads for more common conditions, the use of key workers to promote continuity of care and multidisciplinary teams with specialist expertise, clear links between community and hospital based teams, with essential roles for specialist nurses and GPs with Special Interests (Royal College of Physicians 2011).

Concerns about the limited implementation of the National Service Framework were discussed at a House of Commons Committee and recorded in the 'report of session' (House of Commons Committee of Public Accounts 2012). The report of this session drew on the funding appraisal by the National Audit Office which identified a 38% increase in health spending on neurological conditions from £2.1 billion in 2006-7 to £2.9 billion in 2009-10 (National Audit Office 2011). Concerns were discussed in relation to this increase in investment yet continued lack of improvements in service. The report noted failings in implementation due to lack of national leadership, local commissioners not being held accountable for the implementation, and progress not being monitored. The report also recognised that there were insufficient data measuring the effectiveness of services or providing evidence of best practice for people with neurological conditions. The

research reported in this thesis aims to make a contribution to improve this understanding for HD.

Guidance in the UK refers primarily to neurological conditions as a whole (Neurological Commissioning Support 2011, NHS Modernisation Agency 2005). Some documents are more directive for long-term neurological conditions (Royal College of Physicians 2011) and an even smaller group for progressive long-term conditions. However when this is the case Huntington's disease is rarely included and emphasis is primarily on Multiple Sclerosis, Parkinson's disease and Motor Neurone Disease (Neurological Commissioning Support 2010, Seaton 2008, Fitzpatrick et al. 2010, National Audit Office 2011). Indeed the Neuronavigator provided by Neurological Commissioning Support as a web tool designed to help health and social care commissioners plan services is aimed specifically at services for Multiple Sclerosis, Parkinson's disease and Motor Neurone Disease (Neurological Commissioning Support 2012).

The Neurological Commissioning Support agency is working in conjunction with the national charities for these three progressive long-term neurological conditions. It is not clear why the Huntington's Disease Association is not included in this consultation and development. A further case in point is the Quality Standards to be developed by the National Institute of Clinical Excellence. Again these are specifically for Multiple Sclerosis, Parkinson's disease and Motor Neurone Disease (House of Commons Committee of Public Accounts 2012). It is unclear whether HD sits outside the remit of these documents or whether there it is not possible to make a contribution to such guidance due to a lack of information about HD.

Despite the lack of national government produced guidance the Huntington's Disease Association in the UK provides a range of topic based guidance and information about living with the condition as well as standards of care (Huntington's Disease Association 2011) and specific information for physicians (Rosenblatt et al. 2009) and nurses (Stanley unknown year). A recent special edition of the journal '*Neurodegenerative Disease Management*' provides guidance in a number of papers on all aspects of HD including a

family carer perspective, speech and language therapy, physiotherapy, nutrition, oral health and occupational therapy (Brotherton et al. 2012, Cook et al. 2012, Hamilton et al. 2012a, Hamilton et al. 2012b, Quinn and Busse 2012a, Quinn and Busse 2012b, Sabine 2012, Simpson and Rae 2012). These standards of care have been part of ongoing work by the European Huntington's Disease Network (EHDN) to draw together expertise from across Europe to expand and share knowledge of HD.

SERVICE PROVISION

Simpson and Rae (2012) note that evidence for best practice is lacking and suggest that this reflects the disparity of service provision throughout the world. Across the UK specialist HD services seem to have grown in an ad hoc fashion (Simpson and Rae 2012). It is difficult to identify how many, what type and where HD services are located. Data compiled from the Huntington's Disease Association, the EuroHD¹ projects and personal knowledge identifies 26 UK services providing care for those with HD, although this may not be conclusive. There are very few evaluations of services. Anecdotal evidence and case studies provided by Neurological Commissioning Support (2012) suggest services are primarily directed by a health professional with a particular interest in the condition. Some services have arisen from mental health provision (Dipple and Evans 1998) whereas others were established by a Clinical Nurse Specialist (CNS) or interested neurologist (Kenny and Wilson 2012).

Services can be provided in hospital out-patient and/or community settings (Veenhuizen and Tibben 2009). In the UK services are registered as being provided primarily by neurological or genetics out-patient services. Several identify themselves as multidisciplinary but only two classified themselves as

¹ The European Huntington's Disease Network provides a forum for those affect by HD and professionals working in the field to work together across Europe. The network provides an infrastructure for clinical trials, communication and cooperation. Several sites in the UK are registered as taking part in EuroHD clinical studies. <http://www.euro-hd.net/html/network>

being community rather than hospital based. However it is not clear how many services included a clinical nurse specialist or other health or social care professionals who might visit people in their own homes. One of the tasks of the All Party Parliamentary Group on HD, launched in June 2010 is to investigate this inequity of service provision (Stanley 2012). In line with policy documentation the literature reflects several key aspects of quality HD service provision; multidisciplinary team working and specialist knowledge in conjunction with recognition of the importance of family carers, the promotion of self-management and increasingly, a palliative approach.

MULTIDISCIPLINARY WORKING

As there is currently no cure or way to slow the progression of HD (Dawson et al. 2004) the complex symptoms and behaviours which are characteristic of the disease are managed with a range of therapies including speech and language therapy, physiotherapy and diet and nutrition (Kent 2004, Zinzi et al. 2007). There are a number of drugs that can be used to manage symptoms. However there is often a difficult trade-off between the control of symptoms and the side effects of the drugs (Aubeeluck and Wilson 2008).

The rarity of HD means that most non-specialist care providers such as GPs, District Nurses and staff in care homes are likely to have little experience of the type of care patients and their families may need (van Teijlingen et al. 2001, Bourne et al. 2006). With drives to maintain patients in the community longer a significant burden of responsibility is placed on community-based staff to deliver increasingly complex care in the home environment as the disease progresses. There is limited research on the role in-patient facilities play in the care of people with HD. Existing literature relating to the care of those with neurological conditions mainly relates to community service provision (Wilson et al. 2008a), however, again this focus is particularly on people affected by Parkinson's disease (Post et al. 2010) and Multiple Sclerosis (D'Arcy 2005).

For the majority of their illness, if not all, people with HD live at home with their families, making community-based service provision important. People

with HD may require intensive input from rehabilitation and therapy teams at the same time as symptom management and supportive non-clinical care. Hence a multidisciplinary approach incorporating co-ordinated medical, nursing and social care is advocated by the National Service Framework (Department of Health 2005), the Royal College of Nursing (2008) and by many of those publishing research on HD (Aubeeluck and Wilson 2008, Clough and Blockley 2004, Dawson et al. 2004, Kenny and Wilson 2012, Macleod and Formaglio 2004, Oliver and Borasio 2004, Skirton and Glendinning 1997, Veenhuizen and Tibben 2009, Wilson and Seymour 2007). However, variation in symptoms and changes over time, make long-term planning difficult. Loss of cognitive function with disease progression, together with personality changes, which are sometimes profound, and declining bodily control, often result in substantial disruptions and burdens for informal carers; these are all critical factors to be considered in the provision of community-based care (Bourne et al. 2006).

SPECIALIST NURSING INPUT

There is a paucity of literature on nursing roles in HD (Baker et al. 2009). Baker et al.'s (2009) review identified only six papers relating to this area of research. However, there is evidence that specialist knowledge is important in the management of HD (Dawson et al. 2004, Skirton and Glendinning 1997, Kent 2004). The specialist nurse role is also promoted for neurological conditions by the good practice guide developed by the Royal College of Nursing and allied organisations (2008).

The CNS role is poorly defined (Potter and Coey 2003, Robb 2001) and the Royal College of Nursing report (2008) highlights the lack of consistency and shared understanding of the role in long-term neurological conditions. Despite this lack of clarity the report suggests that the CNS often takes on a leadership role, providing consistency in patient care (Aspinal et al. 2012), often in the role of case manager, initiating interventions, supporting self care and co-ordinating multiple disciplines involved in care provision (Skirton 2005, Kenny and Wilson 2012).

Clinical Nurse Specialists who work in the community have been provided for people with both Parkinson's disease (Bell 2004), and Multiple Sclerosis (Swainger 2001) for some years. A recent audit from New Zealand found that a Huntington's disease specialist nurse reduced HD related admissions to a hospital by 54% and the average length of stay was reduced from 4.3 nights to 2.5 nights (Bourke et al. 2012). However, there has been no national roll-out of such roles in any country. There has been no work to specifically evaluate input of specialist nurses for people with HD in the UK.

A recent study by Kenny and Wilson (2012) evaluated a multidisciplinary team for people with HD and identified the CNS as vital, not only in the care management of patients but also the co-ordination of the team. As the patient population for HD was small team members were seconded to work for the HD service approximately one day per week. The evaluation found that as the team members were not physically located in the same place they predominantly worked in a virtual capacity, relying on email and telephone contact between the monthly clinics (Kenny and Wilson 2012). Work to evaluate the CNS role for Multiple Sclerosis and Parkinson's disease suggested that NHS costs were decreased as a result of reduced emergency admissions and urgent out-patient appointments, as well as improvements to rehabilitation, treatment adherence and patients' quality of life (Swainger 2001, Bell 2004).

The CNS often acts as a care co-ordinator, case manager or key worker (Skirton 2005, Skirton and Glendinning 1997). Case management is seen as the top tier in the 'pyramid of care' (Department of Health 2007) and there is increasing evidence of the benefits of such case management in long-term conditions (Russell et al. 2009, Sargent et al. 2007, Metcalfe 2005). As a case manager the CNS can utilise specialist knowledge of the condition, acting as a central point of contact for patients and other members of the multidisciplinary team (Aspinal et al. 2012, Kenny and Wilson 2012, Skirton 2005, Skirton and Glendinning 1997). In many instances they are also seen as best placed to navigate across organisational boundaries such as between primary and secondary services as well as between health, social services and the voluntary and charitable sectors (Aspinal et al. 2012). Long-term involvement provides continuity of care on a

number of levels (Aspinal et al. 2012) and engenders the trust required for care planning. Having been involved in a patient's long-term care, clinical nurse specialists are often in a unique position to provide high quality care to the patient and family at the end of life (Calne and Kumar 2003). Further to their unique skills the CNS can provide flexible access to care through an open referral system and allow for patients to be seen in out-patient clinics or their own homes (Aspinal et al. 2012).

FAMILY CAREGIVERS

Adults with progressive long-term neurological conditions need help if they are to live at home in comfort. Even with professional and specialist input the availability of a family member or friend who can deliver care and help at home is a key factor in determining how well people manage at home, and how long they can remain in their own home as the disease progresses. Smaller family sizes, wider geographic spread and larger numbers of people in full time employment are all challenges to the provision of informal care.

A UK study by Skirton and Glendinning (1997) to assess the care needs of family living with HD in a large rural area of the country, found that family carers preferred the patient to remain at home. However, as symptoms progressed and particularly as psychiatric disturbances developed, family carers struggled to cope with the patient at home. Furthermore, this same study identified a total of 73 unmet needs including: housing, social services contact, financial advice, carer support and input from various key professionals such as physiotherapy, speech and language therapy, and dietetics. One year follow-up revealed that while 56 of these needs had been met, 10 new needs had arisen, of which six had been met. This highlights the constant varying challenges facing each family affected by HD. The role of informal family carers and the substantial disruptions and burdens that can be placed upon them, are therefore concerns (Aubeeluck 2005).

The essential role of family and other informal caregivers are now becoming increasingly recognised. Carers UK estimate that the economic contribution made by informal carers to the UK each year is £119 billion, more than the

cost of all aspects of the NHS (Buckner and Yeandle 2011). As the needs of carers become increasingly recognised so the scope and nature of the care teams must develop to incorporate members who can meet the needs of family carers as well as the patients (Aubeeluck 2005, Aubeeluck and Moskowitz 2008, Aubeeluck et al. 2011, Carers UK 2004, Kaptein et al. 2007, Lowit and van Teijlingen 2005, Kessler 1993, Semple 1995, Skirton and Glendinning 1997, Skirton et al. 2010). An Australian study emphasised the need to provide practical and psychological support to both patients and family carers but noted that this care must be 'flexible, adequately planned and choreographed' (Dawson et al. 2004, p123). Family carers from the UK and US undertaking Skirton et al.'s (2010) questionnaire survey demonstrated concerns about accessing services, the level of necessary knowledge of those providing services, and about finding appropriate and affordable care as well as accessing benefits (Skirton et al. 2010).

SELF-MANAGEMENT

The protracted disease trajectory for those with HD means that they will be involved with health and social care services for years if not decades. As previously noted, it is not feasible or desirable for services to provide constant intervention or supervision to those with HD. Self-management is widely promoted for those with long-term conditions (Corben and Rosen 2005, Expert Patient Programme 2012) and is considered an important aspect of the Chronic Care Model (Wagner 1998) which is drawn upon to inform the UK's own health care framework (Singh and Ham 2006). However, the focus is on chronic conditions and long-term conditions that are stable or intermittent. There is no evidence for the use of self-management in progressive long-term conditions such as HD.

The limited research available suggests that generic programmes like the Expert Patient Programme (2012) are not suitable for those with long-term neurological conditions (Chaplin et al. 2012). Chaplin et al. suggest that lay-led programmes also often fall short as leaders are unable to answer questions around management of the condition that might arise. The authors identify a

call for 'well-designed condition-specific self-management programmes that cover a wide range of skills-teaching using a multidisciplinary approach' (2012, p251). There is some evidence for the value of programmes for Parkinson's disease such as EduPark (Macht et al. 2007) and Multiple Sclerosis (Rae-Grant et al. 2011).

One recent non-randomised study provides evidence of a beneficial education programme for those with HD (Campo et al. 2012). Adapted from a programme designed for Parkinson's disease patients, it incorporated parallel sessions for patients and family carers of eight 90 minute sessions. Based on core coping strategies the programme provided sessions such as taking an active role in treatment, seeking information, self-monitoring, stress management, active rather than passive coping styles and asking for social support. The effects were assessed before and after delivery of the programme through quantitative quality of life tools and an evaluation on completion. The study found that after participating in the programme those with HD had a significant improvement in behavioural symptoms and anxiety, and family carers reported less psychosocial burden.

Feeding back into services via open access as and when necessary has been identified as important in the literature. In Skirton and Glendinning's (1997) study patients and family carers were told to contact services when they felt these were needed but some were reluctant to do so. They also doubted the quality of the services which they did use (Skirton et al. 2010). Self-management is likely to be best suited to those in the early stages of the condition, who continue to live independently with minimal support from family carers. There is no research designed to explore living with HD in the early stages and it is hoped that this study might be able to contribute in some way to this limited knowledge.

PALLIATIVE CARE

There is now a growing body of literature recognising a palliative care approach for people with HD (Soltysiak et al. 2008, Soltysiak 2008, Travers et al. 2007, Moskowitz and Marder 2001, Kristjanson et al. 2003, Dawson et al.

2004), although only a very small body of literature exists relating to end of life care (Travers et al. 2007, Lindblad et al. 2010, Klager et al. 2008)

Palliative care comes under the umbrella term of supportive care (NICE 2004, p18). It embraces the 'holistic' approach to health care and focuses on quality of life, emphasising open communication, autonomy and choice. The World Health Organisation has adopted an explicitly public health orientation to palliative care, promoting a broad vision as relevant to all those with chronic illness and their families (Sepulveda 2002). The National Service Framework for Long-term Conditions (Department of Health 2005) provides quality requirements for services, of which requirement 9 is of particular relevance in its proposition that people with progressive long-term neurological conditions receive a 'comprehensive range of palliative care services when they need them...' and that these services should meet their needs for '...personal, social psychological and spiritual support, in line with the principles of palliative care' (Department of Health 2005, p5).

There are a range of developments in England aimed at supporting and improving the provision of palliative care for all those in need, shifting from what has been a strong focus on cancer. A key aim of the 'End of Life Care Strategy' (Department of Health 2008), is to provide strategic guidance to improve the care of all those with palliative and end of life care needs in England.

Palliative care needs among people with HD may be present from diagnosis (Travers et al. 2007). As the length of the disease trajectory may be up to 30 years (Soltysiak et al. 2008), professionals may find it difficult to judge when to introduce a palliative approach (Wollin et al. 2006). In its advanced stages HD presents a range of problems which may make both care delivery and advance planning for care in the last stages of life both complex and challenging: ethically, practically and clinically (Wilson and Seymour 2007). This is due to the range of physical, cognitive and emotional problems that people with HD experience as they approach the end of their lives. There can be issues of communication, decision making about nutrition and

technologies of life support, and family care (Huntington's Disease Society of America 1999), making long-term care planning difficult.

CONCLUSION

People with HD can be affected by motor, cognitive and behavioural symptoms and require multifaceted and multidisciplinary input. The genetic nature of the condition means that the implications for families can be far reaching. Many are challenged by how to cope with the knowledge of HD in their family and whether or not to share this information with other family members who may be at risk. The longevity of the condition means that care is primarily delivered in the community, particularly in patient's homes, making the role of family carers essential. However, such caregiving roles can be detrimental to the family members' work, social, psychological and physical well being.

The neurological conditions have been identified as an increasing issue for health services worldwide. There have been concerns that targets provided by guidance for long-term conditions in the UK are not being met despite large amounts of money being invested. This is thought to be due to a lack of leadership and infrastructure in the UK, such as a lack of neurologists. Hence there is no national guidance for HD although the Huntington's Disease Association does provide topic-based guidance and information for service providers as well as patients and family carers.

Research is primarily focused on the search for a cure and much is being done to better understand the way in which the mutated gene affects the brain. The limited research on care delivery supports a multidisciplinary approach, although there have been few studies that evaluate services and little is known about what it is like to live with HD. The next chapter describes the methodological underpinnings of the study and the methods of data collection.

CHAPTER THREE: METHODOLOGY AND METHODS

INTRODUCTION

This study aims to build an understanding of living with HD from the multiple perspectives of people with HD, their family members and the health and social care professional involved in their care. An ethnographic approach was employed to generate data from 15 case studies involving a total of 33 individual participants. The responsive interview technique (Rubin and Rubin 2005) was used alongside observations and visual cues in the form of eco-maps, genograms, physical spaces (e.g. people's homes, clinical settings) and artefacts (e.g. photographs, ornaments, equipment). Participants were recruited from three sites: a community-based service, an out-patient clinic and three residential care centres.

STUDY OBJECTIVES

To gain a holistic understanding of living with, caring for and working in the field of HD to:

- identify the care needs of those diagnosed with HD
- develop understanding of the needs of family caregivers caring for a person with HD
- improve understanding of the issues encountered by health and social care professionals in delivering care to people with HD
- gain an understanding of how care needs change over time
- develop understanding of care provision for those living in the family home and those in residential care settings.

This study contributes to the evidence base for HD by providing a unique focus on people with HD, their family carers and the health and social care professionals involved in their care over a sustained three-year period. By incorporating these three perspectives over time it is possible to build a

comprehensive picture of the needs and experiences of those both receiving and providing care. Particular emphasis is placed on interactions with health and social care professionals. Hence the narratives of health and social care professionals are also incorporated and consultations observed. Schatzki (2005) suggests that to fully understand and grasp 'the point' of activities and interactions we need to understand the person's life more generally so that we may position their actions within wider behaviour and relate it to other phenomena. A case study approach allows the utilisation of a range of methods of data generation over time to gain differing perspectives (Kendall et al. 2009) and directly observe interactive events in order to create a contextual understanding of living with HD.

To familiarise the reader with the nature of the research the first section in this chapter outlines the study design before focusing on the methodological approach adopted. By drawing on elements of grounded theory and ethnography an interpretive constructivist standpoint is discussed. The processes of approvals and recruitment for the study are outlined and the methods of data collection are then described. The chapter then attends to the ethical issues that arose throughout the study. Finally the methods of analysis are presented to address how these data were constructed and interpreted.

STUDY DESIGN

CASE STUDIES

There are so many variations and dimensions to living while affected by HD that this collective case study (Stake 1995) approach was specifically devised to accommodate different family and caring units whether they incorporated family members (with or without HD), friends, or health or social care professionals. The case studies were designed to pursue a network of key people involved in care provision so that a range of opportunities for data collection may be followed in order to achieve in-depth data for each case.

Stake (2000) specifies three types of case study, as shown in Table 1 below. This study uses a collective case study approach to gain a better understanding of a group of cases in order to develop theory which might be applied to a larger still collection of cases (Stake 2000).

Table 1 - Types of case study

Intrinsic case study	Interest is in the stories of the case alone and does not wish to apply this information to a wider context or phenomena.
Instrumental case study	The case is still looked at in depth but its role is to facilitate understandings of something wider.
Collective case studies	Based on the instrumental case study, the concept is applied to several cases. The collective cases may share common characteristics or may be uniquely individual.

The literature on case studies highlights definition of the ‘case’ as a key element of the approach (Bergen and While 2000, Appleton 2002, Yin 2003, Stake 1995). There is debate as to whether cases should be either evolutionary i.e. developing from naturally occurring situations, or selected by the researcher to specifically create the ideal case (Bergen and While 2000). For Robson (1997) there is overlap between these ‘emergent’ and strictly ‘pre-structured’ cases (p148), making flexibility one of the great strengths of the case study approach. The cases in this study reflect this overlap. Despite being predominately emergent in nature in that they integrated ‘naturally occurring’ (Hammersley 1995) family and health care interactions, they also incorporated pre-structured elements. Participants were necessarily identified by staff involved in their care, and bound by limiting the cases to those who were considered ‘key’ carers (formal or informal) by the person with HD (Creswell 1998).

Central to the case is the person with HD. Yin (2003) identifies this type of case study as embedded, using a unit (the person with HD) and several subunits (key family members and health and social care professionals) to

make up one case. In this study a typical case is shown in Figure 3 and consists of:

- the person with HD
- one or more key family caregivers (informal), this was predominately a spouse, although also included a parent, adult children, and one sibling.
- one or more significant health or social care professional. This was predominately the Clinical Nurse Specialist (CNS) for HD, or the Regional Care Advisor (RCA) for the Huntington's Disease Association.

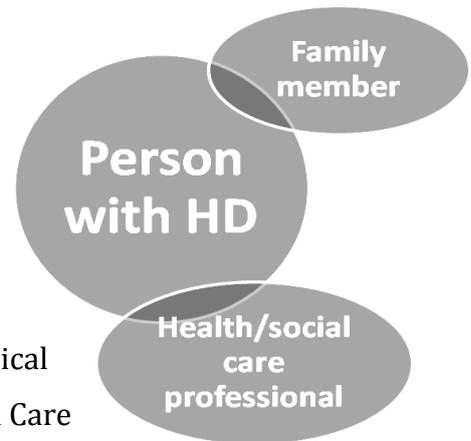


Figure 3 - The case

The emergent and flexible nature of the case study approach meant that it was not possible to predict exactly who was going to be involved as 'sub-units' of each case, particularly as health and social care staff moved in and out of people's lives over time². Using a longitudinal design, each person was contacted at least once a year to ask them to take part in an interview, over a period of up to three years. The frequency of follow-up depended on the stage of the person's disease and stability of their care environment. Observations were undertaken as and when possible and appropriate, focusing on the input of care services.

SETTING

The research predominantly took place in an area of England where two different types of NHS out-patient services situated in adjacent counties could be included. The names of all the sites have been changed in order to maintain the anonymity of the staff as far as possible. Three neurological care centres run by the charity Larch Tree Care Homes were also included to gain perspectives from those using and providing residential services. The four cases living at Larch Tree care centres had already been recruited to a

² See case profiles in Chapter Five for the makeup of individual cases.

previous study providing a pragmatic reason to continue to include these participants. Cases were recruited from three sites which provided services to people affected by HD in different locations, formats and with different professional involvement.

- Birchtree City – a multidisciplinary community team, led by a Clinical Nurse Specialist (CNS) for Huntington’s disease, including a consultant, dietician and speech and language therapist
- Oakfield Town – a clinic based consultant led out-patient service with input from the Regional Care Advisor (RCA) for the Huntington’s Disease Association
- Larch Tree Care Homes – residential long-term nurse-led specialist care (three centres located across England).

SAMPLE

As HD is a relatively rare condition the sample was identified pragmatically based on the knowledge that there was only a small population of people with HD across the recruitment sites. A range of settings was sought in order to compare the different service provision. At the time of recruitment the Birchtree City team held a caseload of approximately 60 people diagnosed or at risk of HD in the area. Across Oakfield Town the caseload for the out-patient clinic was approximately 40 people. The Larch Tree neurological care centres provided residential care for people with a wide range of neurological conditions including HD. Each care centre only had a small number of residents with HD, and the need for the people with HD to have capacity to participate restricted the eligible population to a very limited number.

It was also not possible to know in advance of recruitment of the person with HD how many family members or health and social care professionals might be involved for each case. The aim was for five participants with HD at each site. A sample of 15 cases was identified and achieved.

METHODOLOGY: UNDERSTANDING THE APPROACH

There is much debate about the use of theory in qualitative research (Avis 2003, Reeves et al. 2008, Kelly 2010, Wu and Volker 2009). Reeves et al (2008) argue that theory supplies the foundations for any research study, providing ways of understanding 'things that cannot be pinned down' and giving researchers

" ... different 'lenses' through which to look at complicated problems and social issues, focusing their attention on different aspects of the data and providing a framework within which to conduct their analysis"
(Reeves et al. 2008, p631).

The type of theory used is informed by the perspective of the researcher. This perspective is made up of their world view and fundamental belief systems.

INTERPRETIVE CONSTRUCTIONIST THEORY

This research study was guided by an interpretive constructionist theory. Ontological perspectives are concerned with the nature and experience of being (Guba and Lincoln 1994). This research is shaped by an ontological perspective that rejects the Cartesian mind body dualism and is informed by the conjecture that mind and body are intertwined to constitute a human being (Bleicher 1982). Thus HD is not considered simply as a biological condition but has social, psychological and spiritual implications for the person diagnosed and those around them. Epistemological perspectives are then concerned with what can be known about living with such a condition. Locke (2001) suggests that all ideas are derived from experience. It is the experience of living with HD that gives patients and their families knowledge which can be shared with a researcher to help them understand their 'lived experience'.

Interpretive theory focuses on the person's interpretation of their situation and environment rather than the objective reality of the situation itself, hence behaviour occurs as a response to the subjective interpretation of that

environment. Interpretists take the stance that there can be no objective neutral knowledge as all acts are open to interpretation. However, the theory guides us to highlight elements of shared meaning. Constructivism recognises the co-construction of meaning by all participants in any situation. Each participant constructs their knowledge as a result of their perspective giving it meaning in context, these multiple and sometimes even conflicting accounts can all be valid at the same time (Rubin and Rubin 2005).

The interpretive constructionist standpoint is informed by that outlined by Rubin and Rubin (2005) and Charmaz's construction of grounded theory in which she states that it is not possible to give an exact picture of the studied world but an 'interpretive portrayal' of that world (2006). The work of Diaz Andrade (2009) is also drawn upon in linking interpretive research and the case study approach.

For Guba and Lincoln (1994) a researcher's epistemology requires them to address the nature of the relationship between the 'knower or would be knower and what can be known' (p108). A key issue is whether it is possible for me, as a researcher who does not have HD or have HD in her family, to understand what it is like for a person to live with the disease. As Benton and Craib summarise, the argument raises the question of 'whether we can translate from one form of life to another in a meaningful way' (2001, p98). Bell acknowledges that there are multiple differences with and between cultures but suggests there are fewer 'mysteries in this process' than is often supposed by philosophers (2002, p2). The role of the researcher is to be a vessel to gather and re-present this experiential information in a form which policy makers, and health and social care providers can gain a better understanding of what it is like to live with HD. The ultimate aim is for this information to be used to improve services and supportive practices.

Rubin and Rubin embrace interpretive constructionist theory stating that for a researcher from this standpoint 'how people view an object or event and the meaning that they attribute to it is what is important' (2005, p27). Even though each person understands and interprets an event in an individual way

it is likely that they will also incorporate shared understandings held by family, peers, or members of groups to which that person belongs. For example a home visit by the Clinical Nurse Specialist (CNS) for HD may be interpreted differently by the nurse, the patient and the family caregiver of that patient. However, they may draw on shared meanings of the disease, symptoms, care giving, and family. The researcher must then also be reflexive to recognise her own perspective and cultural assumptions. Attempts to manage assumptions should be made so that they do not influence or restrict the meaning offered by respondents;

“The ability to get into the world of someone who does not share one’s own lenses requires an ability to first recognize and then suspend one’s own cultural assumptions long enough to see and understand another’s”
(Rubin and Rubin 2005, p29).

CASE STUDY

The research employed a case study design in order to gain a robust view of patient and family experiences of living with Huntington’s disease. This is in line with Stake’s collective case study approach incorporating ‘instrumental’ cases which are utilised to provide insight into the care needs of those affected by HD (Stake 1995, Stake 2000, Stake 2006). Cases studies provide a structured approach to collecting research data rather than a particular method (Yin 2003, Stake 2000) and definitions of what constitutes a case study varies greatly. A definition provided by Creswell most appropriately clarifies the concept for the purposes of this study:

“An exploration of a ‘bounded system’ or a case (multiple cases) over time through detailed, in-depth data collection involving multiple sources of information rich in context” (Creswell 1998, p61).

This case study approach was derived from the need to ‘understand complex social phenomena’ (Yin 2003, p2), allowing the exploration of the meaning of experiences and opinions within the context of the real-life events being studied (Yin 2003). Case studies have advanced beyond Yin’s (2003)

positivistic standpoint and increasingly utilise qualitative research designs. Diaz Andrade suggests that this is due to an intersection with grounded theory, which has also evolved beyond its positivist roots (Glaser and Strauss 1967) to an interpretive stance (Charmaz 2006). The fundamental advantage of the qualitative case study for health research is that it can address the complexity of health care situations (Hewitt-Taylor 2002). A case study allows investigation using a range of methods of data generation, often over a period of time, particularly looking at more than one element to establish context and meaning in a more complete way.

THE RESPONSIVE INTERVIEW TECHNIQUE

The case study approach allows the incorporation of a number of people to contribute to the case from differing perspectives. Interviewing taps into these differing perspectives to create a picture of living with HD. As Rubin and Rubin observe;

“Though ordinary life roots you in one position, when you are interviewing, you see life in the round, from all angles, including multiple sides of a dispute and different versions of the same incident. Observing life from separate yet overlapping angles makes the researcher more hesitant to leap to conclusions and encourages more nuanced analysis” (2005, p4).

The present study has embraced this co-construction approach and adopts a model of responsive interviewing. Responsive interviewing as described by Rubin and Rubin (2005) is a model of qualitative interviewing drawing on the interpretive constructionist philosophy with the goal of generating depth rather than breadth of understanding. It remains flexible and recognises the interviewee and interviewer as human beings participating in a particular type of relationship. The case study approach allows versions to be generated from the person with HD, their family member and the health and social care professionals involved (Kendall et al. 2009). The researcher is the ‘tool of discovery’ (Rubin and Rubin 2005, p37), who must be willing to hear what is said and adapt accordingly. Within this model of qualitative interviews the

questions to be asked may only emerge throughout the course of the research.

“Each...new discovery may require a redesign, figuring anew whom to talk to, where to carry out the study, or what concepts and themes to focus on” (Rubin and Rubin 2005, p35).

There are four elements of the responsive interviewing model that are particularly relevant for this piece of research. These are captured in Table 2 below depicting each element of the model and its application to the study.

Table 2 - The responsive interviewing model

	Element of responsive interview model	Application to the study
1.	Learn about what people think about their experiences. Need to involve particular people with certain experiences.	Involved a specific set of participants; people with HD and their nominated family carer, linked to specialist service provision, who wished to, and were able, to share their experiences.
2.	Undertake a number of interviews over time. Topics and questions can be asked and changed based on the responses given.	Allowed follow-up on areas of interest specific for each interviewee.
3.	‘Pause for reflection’ (p37) Recognises analysis as an ongoing process rather than a designated period at the end of data collection.	Reviewed previous interviews, identified areas of follow-up or expansion, and adapted questions accordingly
4.	The emphasis on individual interpretation	Interviewees encouraged to give examples, tell stories and narratives Drew together the differing angles to construct their own picture and understanding of the events.

OBSERVATION

The method of observation is tightly linked with ethnographic enquiry and the concept of participation and immersion in a setting. It is less easy to identify discussion about observation outside of this tradition of ethnography. A comprehensive evaluation of the reasons for using observation as a method for generating data is provided by Mason (2005). The author draws observation and participant observation away from their connections to a particular overall approach and displays them as methods in their own right. Initially it was important to address what observation could add to the case study approach. In discussing this it is possible to address the utility of the data gathered.

The use of observational methods 'usually coincides with the view that *social explanations and arguments* require depth, complexity, roundedness and multidimensionality in data' (Mason 2005, p86, emphasis original). These data are generated in situ and in the context of the setting rather than in a recounted, contrived or restricted way. In this study the observations were able to generate data that was not accessible in other forms and complemented the responsive interview method and visual imagery. Also in line with Mason's (2005) suggested reasons for using observational methods, it could be considered that becoming involved in the lived world of each of the cases may be more ethical than the researcher remaining 'outside' and simply expecting information to be given to them. It was also a way of spending time with the participants without expecting them to take part in a more formal interview. This demonstrated interest in their lives without always expecting them to give information or have me in their home.

Observation research can be a fluid process, making it difficult to plan what will be important during observation periods, and this may change over time (Mulhall 2003). The observations focused on the interactions between people affected with HD, their family carers and health and social care professionals. However the physical environment of the interaction was also important (Mulhall 2003), as was the way people involved in the interactions

constructed their conversations and directed the course of the interaction. These were all aspects noted during or after these sessions.

ECO-MAPS

Stemming from work of pioneering social workers Ann Hartman and Carol Meyer in the 1970s eco-maps were initially used to represent the family's contact with others. As Hartman explains;

“Social workers, in attempting to understand their traditional unit of attention – the person in his total life space over time- are faced with an overwhelming amount of data” (Hartman 1995, p112)

The eco-map draws on the science of ecology, a metaphor that highlights the balance between living things and their environments, which Hartman states can help social workers to see the client as part of a 'complex ecological system' (1995, p113). In this study this is also their purpose. They were used as a diagrammatic representation of the relationships a person with HD has and how these might change over time.

From its origins in social work a small number of nurse researchers have also identified and adapted the eco-map (Keady et al. 2004, Olsen et al. 2004, Ray and Street 2005a, Early et al. 2000). Olsen et al. (2004) argue that integrating the central concepts of pedigrees, genograms and eco-maps is important for holistic nursing. Keady et al.'s (2004) work demonstrates the use of the more traditional eco-map as supporting visual representation in a single case study in order to summarise 'complex issues and relationship-based dynamics' (2004, p60). For Keady et al. (2004), as with those participating in Ray and Street's (2005b) work, the eco-maps were drawn either by or with the help of the participants themselves.

PROCESS OF APPROVALS

Ethical approval was received from the National Research Ethics Service after a substantial amendment was submitted to the Nottingham Committee in June 2007 (see Figure 4 for a summary of the approvals process). The amendment requested the extension of one aspect of a study that was already underway³. Part of the initial study was to use a case study approach to understand the care needs of residents at Larch Tree care centres with long-term progressive neurological conditions. Four of those participants had a diagnosis of HD and were asked to continue their involvement as part of this PhD study. The substantial amendment requested a continuation of this case study aspect by extending the amount of time for a further two years and expanding recruitment to include the Birchtree City and Oakfield Town NHS out-patient services.

A second substantial amendment was submitted in November 2009 for a further extension of 10 months so that as many participants as possible could remain in the study for the targeted three years. Internal ethical approvals were granted by Larch Tree Care Homes. Research and Development approvals were sought from both the relevant Primary Care Trust for the Birchtree site and the Hospital Trust for the Oakfield site. Approval for the Birchtree site was granted in July 2007 and the Oakfield site in September 2007. Both Trusts requested and received annual updates on the progress of the study and a completion report when field work was finished in July 2011.

¹ I had already been involved in the initial ethics submission for the larger project and attended the ethics committee meeting to answer queries. I was also the lead researcher on the study.

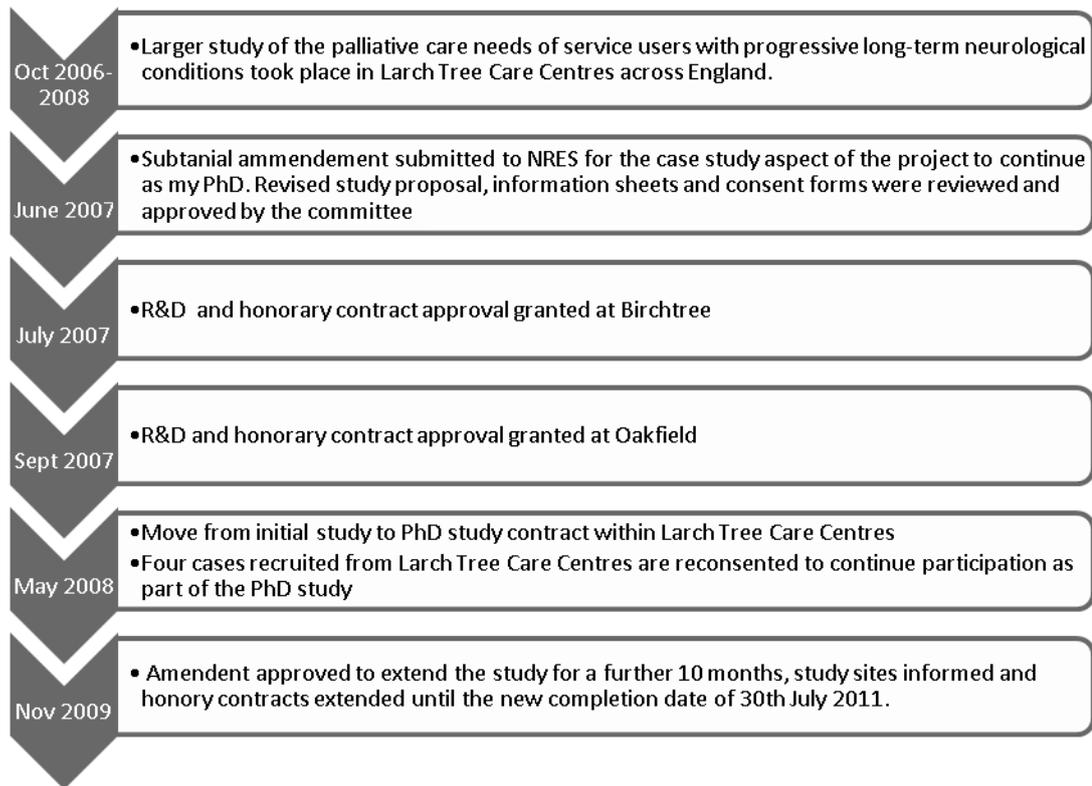


Figure 4 - Summary of the approvals process

RECRUITMENT PROCESS

IDENTIFICATION OF PARTICIPANTS

Links were made with key health and social care professionals within the services at each of the study sites. This support was vital to access service users and their family members as well as the health and social care professionals themselves. People with HD were identified as suitable by the health care professionals working closely with them. In Birchtree this was the Clinical Nurse Specialist (CNS) for HD in consultation with other clinic staff and the consultant for the clinic. In Oakfield it was the Regional Care Advisor (RCA) for the Huntington's Disease Association with the consultant, and at each Larch Tree Care Homes neurological centre, a lead nurse.

In order to take part in the study the person with HD needed to understand the study purpose and have been able to give written consent in some form. The healthcare professionals working closely with the person and their

family had the best knowledge of their abilities and understanding and were used as important gatekeepers to potential participants.

APPROACH

The healthcare professional approached the person with HD and their family member (where appropriate) to explain the study and ask if they would be willing to discuss the study further. It is recognised that this approach will introduce an element of bias. Health and social care professionals effectively selected the potential participants to be approached, and only those known to the clinical team could be approached in this way.

Participants from the two community-based sites were approached during their regular appointments to the out-patient clinics. If the person with HD and their family member indicated they were interested in the research study they were introduced to me and I explained the study further and offered the opportunity to ask questions. Both the person with HD (Appendix B) and their family member (Appendix D) were then given an information sheet to take home and the option to leave a telephone number so that they could be contacted or to make contact themselves if they wished to participate.

RECRUITMENT

When people indicated they were happy to be contacted and left their telephone number I then telephoned them approximately a week later to see if they were still willing to participate and if so to arrange a time to visit. On the day of the visit I telephoned again to check it was still suitable to visit. At this point they could continue with the visit, rearrange the meeting if that time was no longer suitable or change their minds and decide not to take part in the study (see Figure 5 for a diagrammatic representation of this process). Written consent (see Appendices C, E and G for consent forms) was obtained at the first visit prior to starting the initial interview with the participants. From that point onwards consent was verbally renegotiated each time contact was made (process/continuous consent) with each participant prior to undertaking any further interviews or observations.

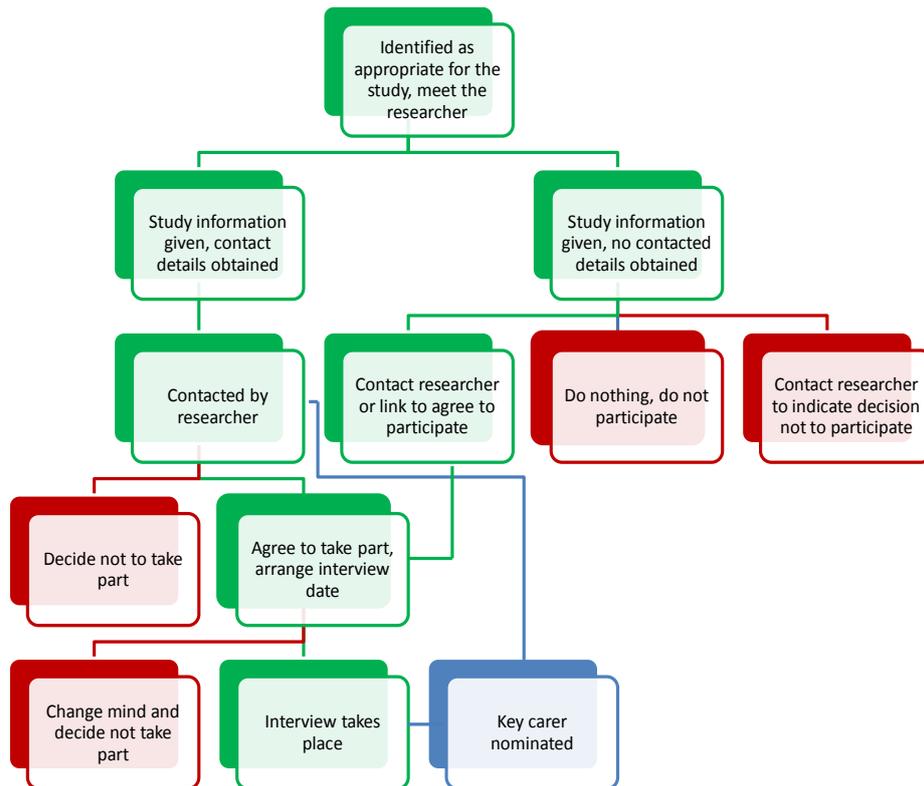


Figure 5 - Recruitment process

RECRUITING HEALTH AND SOCIAL CARE PROFESSIONALS

Participants with HD and family members, where present, were asked to identify a key health or social care professional they had contact with. This was often the person who introduced them to the study, but also included other clinic personnel, a lead nurse, their support worker, or other health professionals who visited them at home. This identified professional was then sent or given written information (Appendix F) about the study with a covering letter to explain why they were being approached. This was followed up with a telephone call to establish if the health or social care professional wished to be involved. No professionals refused. However, if they had indicated that they did not want to be involved they would not have been contacted again regarding the study. Having agreed to take part in the study, participants had the opportunity to ask any further questions they wished and a time and date was arranged for an interview. Opportunities to

extend the network by adding family members or professionals to the case were taken as and when they were available and relevant.

METHODS OF DATA COLLECTION

INTERVIEWS

This study employed the use of interview techniques towards the more unstructured end of the scale, in line with the responsive interview technique (Rubin and Rubin 2005). First interviews were more structured in order to create baseline data which could be used for cross case comparison (Bryman 2001). These first interviews were also used as a platform to build rapport with the respondents. It was recognised that early in the research/participant relationship stories may be recounted in more socially acceptable ways creating a 'public' account (Cornwell 1984). Hence these interviews focused on the participants' HD history and descriptions of more factual aspects of their lives. As a relationship was built over successive visits and interactions it became apparent that more 'private' accounts were surfacing as participants were able to share deeper emotions and describe more complex care and family situations. Subsequent interviews therefore, took a less structured form prompted by identified aspects from previous encounters which warranted further exploration or follow-up (Murray et al. 2009, Rubin and Rubin 2005). These aspects were identified during an initial analysis of the previous interaction. It might be something that had been missed or not fully explored in a previous interview, something viewed in an observation to ask their thoughts about, or perhaps to follow-up on the outcome of an appointment, previously pending interaction with a health professional or a delivery of equipment.

Prompts were used throughout the interviews to encourage the respondent to think more deeply about a particular topic or to think in a way that they had not before in order to gain a more detailed response (Bryman 2001). Prompts, like other questions, were not leading but sometimes used

summary interpretations of the issue or observations to encourage further discussion.

Health and social care professionals were encouraged to draw on the cases known to be included in the study. As an alternative to direct questions in interviews (Rubin and Rubin 2005) these interviewees used the people in the case studies as examples of common features of their work and more unique instances similar to the use of critical incidents and vignettes as identified by Green and Thorogood (2004, p99). Interviews with health and social care professionals were undertaken at their places of work. All interviews with patients and family members took place in their own homes or in the residential care centres.

Joint interviews

The majority of interviews with family members took place as a joint interview with the person with HD. On most occasions it was both the person with HD and their family carers' preference to be interviewed together. This approach also seemed more respectful of the close relationships many couples displayed. It must be acknowledged that by interviewing people with HD and their family carers together a different discussion may have taken place than if each person had been interviewed separately. However, these joint interviews may have encouraged greater openness through the dynamic of the interaction between the participants. Family caregivers were often essential to act as a 'translator' for those with impaired speech and prompt for impaired memory (Shakespeare 1993). Inevitably this often led to the family carer as the more able speaker taking a lead in the interview. This person would often assist the interview by directing their own questions to the person with HD to gain their insight and request that they tell their own story. Couples in particular also displayed co-construction of the conversation by completing each other sentences and contributing prompts. These were often outside the norms of conversational turn taking but helped produce a coherent talk (Bloch and Beeke 2008).

Joint interviews often sparked debate between couples and many aired their differing views as well as confirming each other's perspectives. It could also be suggested that being together allowed both participants to feel more comfortable, particularly when the person with HD might be conscious of their impaired speech or cognition. It also prevented any concerns about what one might say about the other. I felt this was important in order to allow families to feel they could engage with the study in the way that they wished to do so and that respected their family unit.

Some family members did take advantage of when the person with HD left the room to tell me things they obviously did not want to say in front of their loved one. For example when I visited David at his residential care centre it was nearing lunch time and during the interview he left to have his lunch. His wife, Jill and I continued to talk and she became increasingly candid about her feeling towards her husband. As soon as he returned to the room she reverted to much more generic conversation. On two occasions I suspected Tom's wife Nicola deliberately arranged to be out of the house for at least part of the interview to allow Tom time to speak to me alone. She would then join the conversation when she arrived home. Towards the end of the study, once I had a clearer understanding of family dynamics and trust had been built, efforts were made to talk to family members individually where possible. This was achieved for five (Sarah, Martin, David, Sophie, Vicky) of the ten cases where family members were involved. For the other five cases (Harry, Amy, Rose, Tom, Elizabeth) it was not possible to speak to their family caregivers alone. This was because the person with HD was always with them or because they did not wish to participate on their own.

Telephone interviews

As the study continued it became apparent that the continuing and extensive contact with health professionals placed demands on their already limited time. In order to remain sensitive to this and flexible within the case study design telephone interviews were carried out with the CNS in Birchtree and the RCA in Oakfield. Both participated in an in-depth face-to-face interview

early in the study. After this point (in the final two years of the study) telephone interviews were utilised as the most appropriate way of gaining regular up-to-date discussion in relation to specific participants. It is recognised that telephone interviews may not yield the same kinds of data that may be created during a face-to-face interview. However, I felt that any loss of visual cues (Novick 2008) was less important with these health professionals than it might have been with other participants. Telephone interviews provided a pragmatic way to maintain contact with these participants. CNS and RCA were the key contacts at the Birchtree and Oakfield sites making them fundamental to accessing people with HD, their families, clinics and other health professionals. This less intrusive method helped maintain good relations and minimise any feeling that taking part in the research might be arduous or overly time consuming.

Telephone interviews were recorded with consent and transcribed as with all other interviews. They were often shorter and had the advantage of being more direct than the face-to-face interviews conducted. I felt that both these participants remained candid and open with me and both requested the telephone method when the option was given. I think this allowed them an immediacy that was not possible with face-to-face interviews. I would suggest that for busy health professionals having the option of putting one less thing in the diary and completing it there and then was the main attraction of telephone interviews. The limited literature on telephone interviewing in qualitative research, suggests the method is viewed as inferior to that of face-to-face interviews (Novick 2008). I would assert that telephone interviews provide an efficient and valuable method of data collection, and that for health professionals, with whom you have built a good rapport, they can be very effective.

Informal conversations

Throughout this study informal conversations were also utilised. These naturally occurring conversations (Green and Thorogood 2004) were opportunistic and occurred either in the field, for example when attending

out-patient clinics to conduct observations, or during telephone or email conversations with participants. These informal conversations took place with health and social care professionals as well as participants with HD and their family members, and often took the form of brief updates on general health or impending appointments. These contacts were all maintained in field notes in a diary format for each case.

OBSERVATIONS

The interviews were complemented where possible by observational sessions as agreed by the person with HD, their family members, and the health and social care professionals involved. This method allowed flexibility and enabled me to work with the respondents in a sensitive and pragmatic way so that they could engage with the research at a level of involvement they felt comfortable with. No intimate care was observed and observations only took place with the agreement of all those in the vicinity of those being observed (including those not recruited to the study).

The flexibility and unpredictability of this study meant that it was not possible to address in advance whether I would take a participant or purely observer role in the observations sessions. I moved in and out of various roles and degrees of each on a participant/observer scale (Pretzlik 1994, Mulhall 2003, Mason 2005). As some observations focused on interactions with health care professionals there was often only a limited extent to which I could be involved. During observation sessions when I was merely a passive observer, such as, at an out-patient clinic appointment, notes on actions, comments and interactions were taken at the time. My own thoughts were then recorded immediately after in order to note my own feelings about the session and my interpretations of interactions (Emerson et al. 1995).

Other sessions were predominately observational but did allow me to raise questions of my own, such as during a home visit from the clinical nurse specialist or speech and language therapist. At these sessions I could ask for clarification if needed either at the time or after the session. Again in these instances notes were taken at the time and my thoughts recorded afterwards.

In sessions where I was further involved in participation I was not able to make notes at the time but did so immediately afterwards. Some of sessions in particular required a more interactive approach. During the arts and crafts, and the physical activity sessions at the Larch Tree care centres, I fully participated in the session alongside my participants at their request. All notes were typed up into a diary format for each case along with any informal contacts or conversations pertaining to that case.

Ongoing analysis was a valuable part of this research. The fluid, flexible nature of the research design meant that observations were reviewed and used to formulate questions for later interviews and direct where, when, and with whom further observations might take place.

ECO-MAPS

In this study the eco-maps were either drawn with the help of the participants, or were based on data revealed in the interview, and subsequently checked and reviewed by the participants.

The method of eco-mapping used in this work particularly draws on the experiences of Ray and Street (2005a, 2005b). A key element was that the eco-maps were time sensitive. Eco-maps were repeated at least once a year in order to identify changes over time. They recorded both the types of relationships the person with HD engaged in, and also the importance of these relationships to them. In order to encompass all significant aspects of the person's environment the person with HD was asked to identify both key family and social aspects of their lives, as well as formal care arrangements including in-patient, community, clinic, home care and day care services. The mapping process was guided by asking the person with HD questions such as 'who or what is important to you at this time?', 'who helps to look after you?', 'who else is involved in your care?'. On repeat visits a copy of the eco-map was shown to the participant, discussed and any necessary updates or changes made. These eco-maps have been used to demonstrate changes over time in the case profiles in Chapter Five.

GENOGRAMS

Simple genograms have been used in this study to map the inheritance of the disease through the family history as far as possible. Once mapped from the information given in the first interview this was then shown to the participant and checked for accuracy. These were also used as visual cues during interviews and visually displayed the family history indicating known, expected and at-risk family members. Individual family genograms are shown in the case profiles in Chapter Five.

CASE DIARIES

In order to maintain all these different forms of data, diaries for each case were created. These tracked all contacts made with or about the case. Notes were made on informal conversations such as phone calls, emails and ad hoc conversations. All field notes on observations and interviews were also included creating a chronological map of events. Also included were recordings of my own thoughts about that case. Noted were potential areas of further questioning, and reminders of issues or topics to be addressed at interviews either with the person with HD, their family caregiver or the health or social care professional involved.

ETHICAL ISSUES

CONSENT

Consent is an important issue in any study. However, people with HD may have reduced or fluctuating capacity and their ability to retain and recall information can be impaired. In order to meet the needs of the people with HD in this study an information sheet (Appendix B) and consent form (Appendix C) were developed. Process (Dewing 2002, 2007) or continuous consent was used throughout the study. Once written consent was gained at the beginning of the study, verbal consent was sought each time the participant was engaged in further involvement in the research. Hence

participants could agree to participate in some aspects and refuse others. For example, a participant may agree to have their consultation with a health professional observed at one time but feel they do not wish for this to happen at another. They could make these decisions at any time without being withdrawn from the study (unless they wished to do so). Information sheets and consents forms for family members and health and social care professionals are provided in Appendices D-G for information.

RECONSIDERING INFORMED CONSENT

Informed consent is a well formulated topic of debate. The rationale and development of the study documents has been described in a paper published in *Clinical Ethics* (Wilson et al. 2010a) and can be found in Appendix A. Recruiting people to take part in research is difficult in itself, and there is much discussion about whether people who are considered 'vulnerable' should be asked to be involved in research. However much of this work is based on issues for people with cancer and those at the end of life (Pessin et al. 2008, Barnett 2001). Long-term progressive neurological conditions pose an additional set of issues. Reduced cognitive function, such as impaired concentration, slowed thinking, dysphasia or aphasia affect the language skills required to understand, process and recall information, as well as physical issues such as involuntary or impaired movement may be a barrier for recruitment to research.

In order to meet the particular needs of people with HD and develop an information sheet and consent form that would be appropriate expertise from disciplines such as stroke (Bennett 2007), intellectual and learning disability (Strydom and Hall 2001) and older people (Harris and Dyson 2001) were drawn upon. In light of the information gathered from this small area of research an information sheet (see Appendix B) and consent form (see Appendix C) were developed to facilitate understanding of this study by people with HD. The information sheet and consent form incorporate elements such as large font, highlighted key words, plenty of spacing, and pictures, alongside simple language and short sentences.

The information sheet and consent form were viewed and supported by several different parties including the Head of Care Services for the Huntington's Disease Association, a key author in the field, a Clinical Nurse Specialist for HD, and an HD service user. The information sheet and consent form were also approved by the Ethics Committee. This approach to adaptation was viewed positively by the committee and has been used in further studies since.

CONFIDENTIALITY

In a multifaceted study such as this it was not possible to assure participants that their participation in the study would be confidential. Initial recruitment was reliant on health professionals identifying suitable people to take part and I continued to rely on these health professionals to provide updates of any major changes that occurred in participants' lives during the three year follow-up period. However, all participants were assured that what they disclosed would remain confidential. When there were elements that required further exploration with another member of their case permission to do so was requested. During this study much time was spent developing relationships with participants and I found it common to become privy to information which it may not have been necessary for me to know. I feel it is fundamental for researchers to be able to deal with this form of disclosure and make considered decisions about what to do with such information. This was particularly important for this study as data were generated from a number of sources.

IMPARTIALITY

Impartiality was a methodological challenge stemming from the use of multiple accounts and approaches. It could be challenging to remain neutral and impartial to each individual account of the story. This became particularly testing when observational data provided evidence supporting one account over another. During these times it was important to re-evaluate the interpretive standpoint to recognise the individual interpretations of

situations and events rather than try to draw together an objective reality. It was also important in these instances to recognise my own role in the co-construction of data.

RESPECT FOR EMOTIONAL BOUNDARIES

Respect for emotional boundaries particularly relates to the importance of reading non-verbal cues in order to behave appropriately with each individual participant. It was important to judge how to elicit as much information as possible without forcing people to confront issues or areas they were not comfortable talking about. Sometimes participants would give verbal indications by simply stating that they did not want to think about a certain aspect. The longitudinal aspect of this study allowed a relationship to be built with the individual participants, and was beneficial in reading situations and responses.

PERSONAL AND PROFESSIONAL RISK

Due to the small population of people with HD in the area and in particular the limited number of health and social care professionals regularly involved in their care there are limitations to anonymity and confidentiality. Every effort has been made to maintain anonymity by changing the names of the sites where the research took place, providing all patients and family carers with pseudonyms and only identifying professionals by their job title. In particular there was only one CNS and RCA working in the research areas and it may be possible for them to be identifiable to themselves and others in the field should they read this thesis.

Confidentiality is particularly problematic in case study research as context, extra details and multiple perspectives are important facets of the approach. Each participant gave their consent to participate throughout the three year study period and all knew of the other members involved in their case. Interviews with patients and family members were predominantly undertaken jointly, as was their preference. None of the participants raised concerns about these issues at any point during the study. However some

data has been excluded to maintain the privacy of participants where it has not held relevance and impact for the study. Extracts from two case studies have already been published in book chapters (see list of publications at the start of this thesis). Participants in these cases reviewed and commented on these pieces prior to publication. One participant requested copies of her interview transcripts and these were sent to her.

Open discussion was maintained with all participants about the length of the research study. Over such a long period it is possible that participating in the research may have had an impact on the participants' lives. During their penultimate interview, each person with HD and their family member was informed that their participation was due to complete and each was involved in identifying when approximately this would be. In setting up the final interview date it was again reiterated that this would be the last time that contact would be made as part of the study. As the majority of participants were able to complete the ideal time of three years they constructed this as a proud achievement. After the final interview a 'thank you' card was sent to the participants to thank them for their participation, and to signify the end of the study for them.

ANALYSIS

DATA TRANSFORMATION AND MANAGEMENT

All formal interviews were recorded with the consent of those present. Throughout the study identification codes were used in transcriptions, notes and diaries in order to maintain confidentiality. The length of the interviews varied from person to person and at different times of the study. All interviews were determined by the interviewee's willingness to share their views and on some occasions, their health. All audio recordings were stored on a secure university server and, deleted from the recording device as soon as they were uploaded. I did much of the transcription myself but a service was also used in order to remain up-to-date with transcribing and allow

ongoing analysis of the data. Confidentiality was assured by the transcription service and all files were transferred via a secure internet portal. Once transcripts were received they were reviewed in conjunction with the audio file to check accuracy and revise any sections which may have been difficult to transcribe due to the interviewees' impaired speech.

All transcripts were stored on the university secured server. Hard copies were stored in a locked filing cabinet in a locked office on the university premises. Hard copies of field notes and diaries were also stored in this way. For the purposes of supervision, extracts of transcripts, diaries and field notes were shared with my supervisors, all of which were anonymised. One of my supervisors had conducted research on HD and knew one of the participants well. I spoke with this participant about this and she was happy for this supervisor to read her interview transcripts and view data related to this case where necessary.

CODING

Analysis was ongoing throughout this study and is an integral part of the responsive interview technique (Rubin and Rubin 2005). All data were stored and managed in QSR NVivo8 (QSR International Pty Ltd 2008). Interviews were imported once transcribed and checked, and diaries were maintained directly in the software package. Each transcript was initially individually hand coded and those codes reviewed when coding was repeated electronically using the NVivo software. This allowed rethinking and reorganisation of codes in the light of additional data. All codes were labelled and given a description. To organise data in the computer software use was made of both the 'free node' and 'tree node' functions to sort and rank codes. In many instances it was clear how concepts related to each other so it was possible to build tree nodes, creating a hierarchy of codes. Other codes were less easy to place and free nodes were used to capture these data. Individual or groups of codes were also printed to promote engagement with the content of the code and potential themes that may then be constructed. All coding remained flexible so that codes could easily be moved between

free nodes and tree nodes as well as to different tree nodes, or level of the node. This occurred as data were reanalysed and the importance of different codes ebbed and flowed.

BUILDING CASES

Once data collection was complete for a case all data from that case were reviewed and recoded where needed in order to see each case individually over time. Each case was written up as an individual narrative and edited versions of these are provided in Chapter Five. In addition, interview data for each case was supplemented by observational data and field note entries from case diaries. Genograms and eco-maps were also used to generate context and perspective to interactions. These individual case narratives provided a clearer view of the key elements of each case, their family contacts, living situation, care input and changes over time. Figure 6 shows how the different types of data are intertwined to create detailed context around the views and opinions provide by the verbal narratives of the interviews.

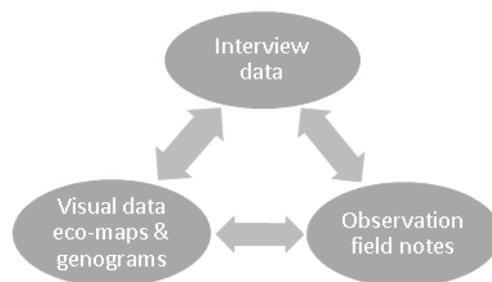


Figure 6 - Interaction of data

It was important to build case narratives in order to be able to view each case individually and remain true to the case study approach. This prevented cases from becoming swamped or 'mangled' by the cross-case analysis and allowed the 'unique vitality' of each case to be displayed within its own context (Stake 2006, p39).

BUILDING THEMES

The individual case was studied to gain understanding of the care needs of those affected by HD as far as that situated case could provide (Stake 2006). However, it was not possible to just regard each case in isolation. Analysis had to take place both within and across cases. Cross case analysis expanded understanding by comparing cases to identify commonalities, concepts, typologies and anomalies. As Stake illustrates, the complex meanings of the study are 'understood differently and better because of the particular activity and contexts of each Case' (2006, p40).

Cross case analysis took place several times throughout the study period and in a number of different ways. In the first instance cases were divided by time period of the interview such as; first, second, third interviews. Then by participant type; people with HD, family members, and health and social care professionals. Later in the process of data collection data sets became more complete and it was then possible to use modelling or spider diagrams to start to group codes into themes and identify relationships between codes. Throughout this process memos were kept in order capture ideas and the direction of coding. As analysis took place over a number of years these memos became essential to understanding the decisions I had made about certain codes and reasons for the content of the text captured in those codes. Figure 7 shows an extract of the tree coding hierarchy for the node 'Discovery'. This node shows three levels of hierarchy and the other nodes used to explore this theme in NVivo.

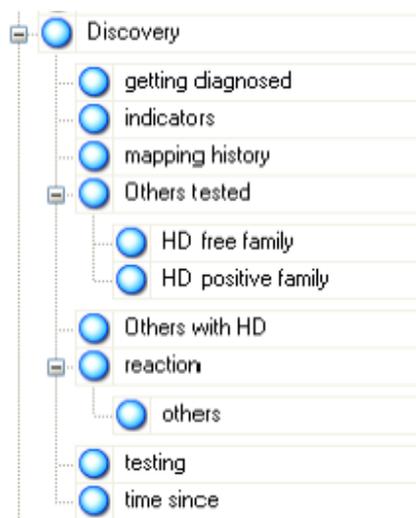


Figure 7 – Example of coding of a tree node

I was aware that by being immersed in a longitudinal qualitative study I would develop a good sense of narratives being constructed by the data. However, as Rubin and Rubin (2005) note, my notion of what might be important in each person's story could be selective. It was important to carry out repeated forms of analysis so as to thoroughly interrogate the data. These approaches also needed to be carried out systematically so as to remain open to any and all data rather than look for confirmation of my initial thoughts. I felt it was not about dismissing my initial ideas but employing caution, as Charmaz suggests, so that my initial ideas represent 'one view among many' (2005, p54). It was in carrying out analysis in this way that I was able to formulate a temporal approach to presenting the narratives in relation to the individual stories told by each case. These 'cross case' and 'within case' analytical formats allow for broader themes to be developed without losing the context of each case.

CHAPTER FOUR: BUILDING CASES

INTRODUCTION

The chapter is divided into three sections. The first gives an overview of the three settings used in this study. The second reports the demographic profile of the sample. In the third part of this chapter I reflect on the impact of talking to people with physical and cognitive impairments for the purposes of research. This section describes the interview process and the adaptations needed to incorporate the voice of the people with HD. I then go on to discuss the rigour of the data collected and how generalisations might be made for the wider theory in relation to HD. This chapter provides an essential background for the individual cases. Demographic information on how and when participants were recruited, the types, frequency and duration of participation and participant's knowledge of HD in the family are presented. This chapter contextualises the data for a clearer understanding of the cross case synthesis of the qualitative data presented in Chapters Six, Seven and Eight.

The names of all persons and the sites of study have been changed to ensure anonymity and confidentiality. Health and social care professionals are referred to only by professional role.

SETTINGS

BIRCHTREE CITY

The HD service in Birchtree city was established in 1994 and was co-ordinated by the CNS. The team consists of the CNS, a consultant neurologist, a speech and language therapist, a dietician and a rehabilitation support worker (who joined the team in April 2009 and works two days per week). The CNS is the only full

time member of the team, the rest of the team work on a secondment⁴ basis providing input approximately one day per week. The CNS provides a single point of contact for the team and service users. The service provides an open referral system so that anyone may refer to the team, including patients and families. People are seen in out-patient clinics. One is based at a hospital and the other at a community health centre. Patients and their families are also seen in their own homes and within residential and day care settings.

OAKFIELD TOWN

In Oakfield town an NHS hospital based out-patient clinic was run by a consultant in rehabilitation medicine. The RCA for the area had strong links to the clinic, built over many years and attended in conjunction with the consultant. Patients and families were seen at the hospital and could be referred in a number of different ways. Via the hospital there was also access to psychiatric services, dietetics, speech and language therapy and physiotherapy. The RCA also visited patients and families at home and within residential and day care settings.

LARCH TREE CARE HOMES

The study involved four residents from three of Larch Tree's specialist neurological care centres. Larch Tree Care Homes is a national charity providing palliative, neurological and home care for people with complex illnesses and conditions. The three care centres varied in size from a 36 to 44 beds. All were located in converted stately homes, two in rural areas and one in a small town centre. The care centres are all nurse-led with personal care provided by support workers⁵ and medical referrals to local GP practices and neurological teams. The care centres offered therapies such as physiotherapy and multi-sensory therapy alongside recreational therapies such as gardening, arts and crafts, games and Pets as Therapy.

⁴ Team members such as the dietician and speech and language therapist work in wider roles in the NHS but are contracted to work a set number of hours per week for the HD service.

⁵ Across the study, care centres used the terms 'Support worker' and 'Health Care Assistant', for the purposes of this thesis and for consistency just 'Support Worker' has been used.

CASE DEMOGRAPHICS

RECRUITMENT

The majority of participants were recruited via out-patient clinics at Birchtown city and Oakfield town. In Birchtown the clinic ran monthly and approximately every two months at Oakfield. Clinics were attended over a two year period until a spread of cases across the sites was achieved. Of the 55 clinic appointments attended by people with HD (this does not account for repeat visits during this time) 17 people were invited to participate in the study. Reasons for not inviting people to participate included: repeat visit, the patient lacked capacity to understand the study, the health professionals involved did not consider them suitable for participation, that it was not a suitable time to ask them to participate, or they were missed. Three of the seventeen invited refused directly, and five indirectly by not responding after taking information home to consider. Nine agreed to take part, six from Birchtown city and three from Oakfield town (see Table 3).

Table 3 - Clinics accessed, number of patients given information and identification of those recruited according to clinic location and date

Location of clinic	Date of clinic	Total no. patients in clinic	Given info sheet	Refused after follow-up	Non-response refusals	Recruited
Birchtown	Sept 2007	5	3	0	2	Sarah
Birchtown	Nov 2007	4	2	0	0	Helen, Rose
Oakfield	Dec 2007	4	4	1	1 (re-invited Vicky)	Amy, Erica
Oakfield	Jan 2008	5	2	1	1	0
Birchtown	Jan 2008	7	2	0	0	Martin, Sophie
Birchtown	Feb 2008	5	1	0	1	0
Oakfield	April 2008	3	0	0	0	0
Oakfield	June 2008	4	0	0	0	0
Birchtown	Sept 2008	5	1	0	0	Tom
Oakfield	Jan 2009	7	1	1	0	0
Oakfield	April 2009	6	1	0	0	Elizabeth
Total	11	55	17	3	5	9

Non-clinic recruitment

Kate, Alison, Mary and David: The four cases based at Larch Tree care centres were recruited as part of an ongoing project with this organisation (Wilson et al. 2008b, Wilson and Seymour 2007). A small aspect of that study involved six case studies, two participants with Multiple Sclerosis and four with HD. The participants with HD were asked to then continue on in my PhD work. Their participation would remain the same, however the shift in use of the data was explained and each was asked to give additional written consent to continue. All were happy to continue being involved, as were their family members. The overlap in the studies was for a period of 11 months between June 2007 and May 2008. All the interviews for both studies were conducted by myself and although interviews undertaken during this time were used for both studies, the earlier study focused on palliative and end of life care needs and did not draw on wider themes from those participants' stories.

Harry: Harry and his wife Margaret volunteered to take part in the study when I was introducing it at a local Family Day organised by the Huntington's Disease Association in June 2007. Once study approvals had been confirmed for Birchtree area I contacted the couple to ask if they still wished to participate. They agreed to do so and I arranged to visit them at home for a first interview.

Vicky: In an attempt to gain a more even spread of participants between the three sites, the Regional Care Advisor in Oakfield identified and approached one person that had been given the information in clinic previously but had not responded. The RCA felt that her situation had become more settled and she would now wish to take part (one year after first meeting her in clinic). I then contacted Vicky to ask if she was willing to participate, and we arranged a time for me to visit her at home for a first interview. During this time I also posted another information sheet to her home.

Recruitment period

Time taken to recruit the cases from each site varied as shown in Table 4. Due to a smaller number of people with HD in the area the clinics in Oakfield ran less frequently than those in Birchtree. More participants could have

been recruited from the Birchtree area to achieve the sample sooner. However, I decided that maintaining as good a spread as possible across the three sites was an important part of the study in order to be able to look at some of the differences between the services at each site and so continued to try to recruit participants from the Oakfield site.

Table 4 - Recruitment period and number of cases recruited per site

Site	Recruitment period	Length of time for recruitment	Number of cases
Larch Tree Care Homes	June 2007 – April 2008	10 months	4
Birchtree City	September 2007 - September 2008	12 months	7
Oakfield Town	December 2007 - January 2009	14 months	4
Total	June 2007 - January 2009	19 months	15

STUDY DURATION

The aim was for all the people with HD to participate in the study for three years. The minimum time a case was involved in the study was 11 months. This was also the only participant to be withdrawn (Mary). Eleven of the 15 cases remained in the study for the full three years. Due to delayed recruitment, the data collection period ended before the last three cases were able to complete three years in the study. These cases completed 2 years 8 months (Tom), 2 years, 7 months (Vicky) and 2 years 3 months (Elizabeth) respectively. Dates and duration of participation are shown in each case profile in the next chapter. Figure 8 also shows each participant's duration in the study and the times when data collection took place.

No one chose to withdraw from the study. Two participants were sectioned under the Mental Health Act and moved to mental health facilities. For one participant this ended her participation in the study (Mary). For the other (Erica) this breakdown in her mental health occurred towards the end of the

study period and staff caring for her deemed her well enough to give consent for me to visit her for a final interview at the mental health unit where she was staying.

In Sarah's case I chose to withdraw from contact approximately 4 months before the end of her three year study period. After discussion with her family and the health professionals involved we all felt her capacity had declined to the point where she was no longer clear about who I was or what I was asking her to be involved in. Her husband wished to remain in the study and completed the three year follow-up period. Another participant, Alison moved to a different care home after her Larch Tree care centre was closed. The nursing manager allowed me to visit one last time to conduct an exit interview. However, lack of access to this additional care home (not within the approvals of this study) prevented further follow-up, making my last contact with Alison at two years nine months.

	2007		2008			2009			2010			2011	
	June-Aug	Sept-Dec	Jan-April	May-Aug	Sept-Dec	Jan-April	May-Aug	Sept-Dec	Jan-April	May-Aug	Sept-Dec	Jan-April	May-Aug
Alison		P	P	P, S		P, S			P				
Amy			J		J		O, S		O	J, S		O, R	
David			J			J			J, O				O, R
Elizabeth						J		O, S		O, J	O		S, J
Erica			P			P, O		S	O		P	O, P	S
Harry	J	O		J	O	O, O, O, S	O		J	O			
Helen	P			O, P		O, S		P		O, P, S			
Kate	P	P		O		P, S			P, O, S				
Martin			J	O		O	S	J	O, O	O, J, O	S	O, R	
Mary			P	O			S						
Rose			P	O	O		S	O	J	O		J, S	
Sarah	J		R, O	O		J	O, S	S	O	R	R		
Sophie				J		J	S		O, O			R	O, R
Tom					J		O, O, S			P/J		O	P/J, S
Vicky					P		O, R	S			P, R	O	R, S

Figure 8 -Case involvement and data collection points over time for each participant

P=Patient interview; R=Relatives interview; J=Joint interview; S=Staff interview; O=Observation
(P/J=Interview started with patient only and then a relative joined)

STUDY PARTICIPATION

The interviews in this study took place over a data collection period of four years, July 2007 to July 2011. They were undertaken with people with HD, members of their family and the health and social care professionals they came into contact with throughout their participation in the study. Table 5 below shows the total number of interviews within each participant category.

Table 5 - Total number of interviews per participant group

Participants	Number of interviews
People with HD alone	21
Family members alone	11
Person with HD & family member together	24
Health and social care professionals	12*
Total number of interviews:	68

*Seven individual health and social care professionals were interviewed, 3 were interviewed twice and 1 was interviewed three times.

In order to maintain flexibility for each case observations varied in length and location. Specific length of time spent on each observation was not recorded but they ranged from approximately 20 minutes to 2 hours. Type and frequency of observations are shown in Table 6.

Table 6 - Observation type and frequency

Type of observation	Frequency
Out-patient clinic appointment	30
Clinical nurse specialist home visit	9
SALT home visit	3
Activities session in residential care	3
Day care	1
Physiotherapy session	1
Total number of observations	47

Cases have been treated individually throughout this study. Each is made up of different participants and the number of interviews, observations and eco-maps varies. These elements are portrayed in the case profiles in the next chapter. Health and social care input came primarily from two sources, the

Clinical Nurse Specialist (CNS) in Birchtree and the Regional Care Advisor (RCA) for Oakfield, hence their interviews are incorporated in more than one case. The number of interviews with health and social care professionals and their relation to each case is shown in Table 7 below.

Table 7 - Interviews with health and social care professionals

Health or Social Care Professional	Number of interviews	Date of interview/s	Participants discussed
CNS	3	24.06.2009	Harry, Sarah, Helen, Martin, Rose, Tom
		14.12.2010	Helen, Amy
		27.06.2011	Sophie, Tom
RCA	2	20.10.2009	Amy, Erica, Vicky, Elizabeth
		27.06.2011	Erica, Vicky, Elizabeth
Staff nurse	2	03.07.2009	Kate, Mary
		29.07.2010	Kate
Support worker	2	18.07.2008	Alison
		19.06.2009	Alison
Staff nurse	1	24.07.2009	Sarah
SALT	1	21.09.2010	Martin
RSW	1	29.03.2011	Rose
Total	12		

CNS=Clinical Nurse Specialist, RCA=Regional Care Advisor, SALT=Speech and Language Therapist, RSW=Rehabilitation Support Worker.

For those in residential care input was sought from two staff nurses and a support worker. As the role of the speech and language therapist (SALT) became increasingly important in Martin's case and the Rehabilitation Support Worker (RSW) in Rose's case, interviews were also conducted with these health professionals to better understand their roles in the respective case.

As Table 8 shows, five cases did not have input from family members. Of the family members involved in the study, seven were spousal relations but other family members also included one sister, one mother and two daughters. No observations were conducted with Alison. She did not undertake any of the activities provided by the residential care centre and I was not able to arrange to see her out of this environment. To compensate, additional interviews were conducted and I was able to speak with her support worker

at the residential care centre. In David's case, no staff members were interviewed although a meeting with residential care centre staff, his wife and the CNS for his locality was observed as well as a discussion between his wife and the CNS. These interactions are reported in Table 8.

Table 8 - Breakdown of interviews and observations by case, type and frequency

Case	Interviews – person with HD	Interviews – family member	Interviews – HSPC	Observations – type and frequency
1	Alison - 5	n/a	Support worker - 2	
2	Amy - 3	Ben (husband) - 4	RCA – 1 CNS - 1	2 - clinic appointment 1 - day care centre visit
3	David - 3	Jill (wife) - 4		1 – physical activities session 1 – care meeting 1 – discussion between the CNS and Jill, David's wife
4	Elizabeth - 3	James (husband) - 3	RCA - 2	3 – clinic appointment
5	Erica - 4	n/a	RCA - 2	3 – clinic appointments
6	Harry - 3	Margaret (wife) – 3	CNS - 1	4 - CNS home visit 3 - clinic appointment
7	Helen - 3	n/a	CNS - 2	3 – clinic appointments
8	Kate - 4	n/a	Staff nurse - 2	1 - Arts and craft session 1 – Tea party
9	Martin - 3	Julia (wife) – 4	CNS – 1 SALT - 1	1 – CNS home visit 2 - SALT home visit 4 – clinic appointments
10	Mary - 1	n/a	Staff nurse - 1	1 – physiotherapy session
11	Rose - 3	Charlotte (daughter) - 2	CNS – 1 RSW - 1	1 - CNS home visit 3 – clinic appointments
12	Sarah - 2	Max (husband) - 5	CNS - 1 Staff nurse – 1	1 - care home visit 3 - Clinic appointment
13	Sophie - 2	Melanie (sister) - 4	CNS - 1	2 – clinic appointment 1 – SALT home visit
14	Tom - 3	Wife Nicola – 3 Daughter Abigail - 1	CNS - 2	1 - CNS home visit 2 – clinic appointment
15	Vicky - 2	Sheila (mother) - 3	RCA - 2	2 – clinic appointment
Total	45 (21 single, 24 joint*)	35 (11 single, 24 joint)	12 (individual interviews)	48 (individual sessions)

*Two of these interviews started with the person with HD on their own, the family member then joined the interview part way through.

CNS=Clinical Nurse Specialist, RCA=Regional Care Advisor, SALT=Speech and Language Therapist, RSW=Rehabilitation Support Worker.

DISEASE OVERVIEW

Each of the participants recounted their route to diagnosis. Table 9 presents each of the participants' age, date of diagnosis and genetic source of the disease. For seven it was the beginning of their lives with HD as they were not aware of HD in their family. Eight participants were aware that HD was in their family. Three (Kate, David and Vicky) had searched for their own diagnosis prior to the presentation of symptoms (although it was not possible to identify the exact onset of symptoms). When investigating the early death of his father in an accident David found recorded symptoms of HD in his father's medical notes. His father had died before an official diagnosis was given but this prompted David to investigate his father's family further and request to be tested himself. Table 9 also shows that for Alison and Elizabeth it is still unclear which of their parents carried the HD gene. Both Elizabeth's parents had died at an early age, suggesting non-penetrance. However both Alison's parents had lived beyond their 80s suggesting a new mutation, non-paternity or a small possibility of non-penetrance.

Table 9 - Knowledge of HD in the family

Name	Age at diagnosis	Date of diagnosis	Time since diagnosis (at start of study)	Knowledge of HD in family	Affected parent
Alison	54	2001 approx	6	No	Unclear
Amy	27	1997	10	Yes	Father
David	41	1993	15	No	Father
Elizabeth	59	2008	1	No	Unclear
Erica	52	2005	3	No	Mother
Harry	66	2004	3	No	Mother
Helen	68	2005	2	Yes	Mother
Kate	30	1995	12	Yes	Mother
Martin	55	2003	5	Yes	Mother
Mary	40	1996	12	Yes	Mother
Rose	60	2007	1	Yes	Mother
Sarah	55	2001	6	No	Father
Sophie	40	2002	7	Yes	Father
Tom	45	2007	1	No	Father
Vicky	19	2002	6	Yes	Father

This section has presented the demographic data from this study in order to give an overview of all the cases together and illustrate some of the differences between the data collected as part of each case. In the next section I reflect on the interviewing process in order to highlight some of the challenges for data collection. The final section will then reflect on the validity of the data collected and how these might represent the participants' stories and a wider understanding of living with HD.

RECONSIDERING QUALITATIVE INTERVIEWING

The complexity of the symptoms of HD clearly raise a number of issues for researchers wishing to understand the experiences of those living with the condition. It is arguable as to whether interviews are a suitable form of data collection for this group of people. However, the nature of HD means that symptoms vary from person to person and over time, hence not all participants in this study had the same level of speech difficulties or cognitive impairments. It was still important to gain the views of those with impairments and reduced abilities. The challenge was for me to adapt my technique to the participant group in order to include them as fully as possible.

“Recognizing that some people may be harder to interview than others, it remains the task of the interviewer to motivate and facilitate the subjects’ accounts and to obtain interviews rich in knowledge from virtually every subject” (Kvale and Brinkmann 2009, p165)

This section outlines the learning from undertaking interviews with people with HD and draws on the cases themselves in order to illustrate the techniques employed to enhance the process.

People with HD present particular challenges to a researcher’s interview skills. As always with HD the learning is ongoing, and has been throughout this study. I have been taught by those who have HD and those who know them most intimately how to adapt my skills in order to work in their world. Again it is important to note that my participants all varied in their abilities and were affected by the triad of symptoms in different ways. Listed below are the elements of HD that I found to affect the interview process most directly:

- Dysphasia/Aphasia - Affecting the language skills required to process and understand information, thus it is not just speech but loss of language.
- Communication –Harder to assimilate the information being given and for the appropriate response. This can be helped by allowing time for the

person to respond, by asking one question or using one sentence at a time.

- Concentration – Easily distracted by noise or interruptions, it is important to minimise these as far as possible.
- Memory problems – Difficulty with taking information in, storing it and particularly retrieving it. Prompts and cues can assist retrieval.
- Involuntary movements – the person with HD may be self conscious of these or they can be disruptive to the interview, or a person maybe tired by trying to contain their movements in ‘public’.

How these challenges to the qualitative interview process were experienced and addressed is now discussed by drawing on examples from the cases in the study.

IMPAIRED SPEECH

Impaired speech was one of the key challenges to address when interviewing people with HD. As Kvale and Brinkmann (2009) identify in their seven features of interview knowledge, knowledge is linguistic (p55), and language is the medium of interview research. The oral interaction is the product of the interview which is transcribed and presented as text. Although not a traditional barrier created by speaking different languages, the speech difficulties and cognitive impairments often apparent in people with HD did create some need for ‘translation’. In these instances health professionals and family members who knew the person well became invaluable in helping translate and interpret things said by the person with HD.

During an interview family carers often fluctuated between being a respondent themselves and acting as a translator for the person with HD. Green and Thorogood (2004) address the use of translators and interpreters in research, identifying their role ‘to translate not just the literal meaning of the words used by respondents but the contextual information also carried’ (p84).

Participants had a range of impairments that hindered smooth discussion. For some speech was just a little slurred or jerky. However, for six

participants speech was greatly impaired and I often needed assistance in understanding what was said. It may be that asking them to repeat was sufficient. At other times having a family caregiver present to translate was extremely beneficial. Bloch and Beeke (2008) suggest natural turn taking in conversation allows talk to be constructed collaboratively; therefore there could be value in considering how talk might be constructed in this way when one party has language difficulties. The authors draw together the relevant literature to provide examples of three types of co-constructed talk, all of which I experienced when undertaking interviews with people with HD. Firstly, instances when response construction is shared so that the person with HD may start the response but it is completed by the other person. In this way the perception that they can complete the utterance remains intact so that they continue to engage with the interview. A second example would be for the person with HD to respond and then the family caregiver 'unpack' (p977) this information and effectively speak on their behalf. This was a common practice as participants with HD would often respond to questions with brief statements and the additional information supplied by family caregivers was essential to understand the context of the answer. A third occurrence might be for the person with HD to give an unclear or incomplete response. The second person would then rearticulate the utterance with what the person with HD meant to say, this would then be confirmed by the person with HD (Bloch and Beeke 2008).

For two participants (Alison and Mary) no family carers were available to assist with translation. In order to minimize the 'language barrier' it was important to get to know the participants over the study period so that I could 'tune my ear' to their speech and emphasis. Stress can also affect speech difficulties, hence getting to know the participants and creating a relaxed atmosphere was essential. Speech impairments often became more pronounced as the person became tired, therefore this was taken in to consideration when arranging interviews.

USING VISUAL CUES

Even when speech is impaired it is not removed, and it may be supplemented by other forms of communication, for example visual cues and imagery (Green and Thorogood 2004). During the interview process visual cues were drawn upon when appropriate to initiate responses and put participants at ease by talking about something familiar to them. This was particularly important during first interviews. As interviews with people with HD and family carers took place in their homes personal objects, such as photographs, pictures and trinkets were often used to encourage people to talk about their family members, places they had been or things they had done. This took place to a greater or lesser extent depending on the respondent. For example Kate liked to show me things in her room at her residential care centre and explain where they had come from. This helped me to understand the things that were important to her, what she enjoyed doing, and who was involved in her life. I was then able to encourage her to express herself in this way in future interviews. She would tell me about the family pictures on her wall, show me cinema and theatre tickets to explain what outing she had been on, as well as medical supplies she stored in her room such as fortified drinks. Participants would also show me adaptations they had made to their houses to accommodate the person with HD at home. Julia showed me the downstairs bedroom, wet room and the doorway they had incorporated to link Martin's room directly to the kitchen. Drawing the eco-maps was also a way to supplement the interview with visual imagery and prompt discussion about the involvement of services.

COGNITIVE IMPAIRMENTS

The cognitive impairments of slowed thinking and difficulty concentrating also posed challenges for conducting the interviews. Qualitative interviewing steers the researcher towards an open question style, responsive to the answers given, and incorporating prompts. The fundamental ethos is to allow the respondent to actively co-construct knowledge by encouraging them to guide the interview topics and express themselves using the language they wish. Talking to people with HD required me to adapt the interview process

in terms of length of interview, management of the interview focus and the way in which questions were phrased.

It was clear that for some participants concentration was often a concerted effort, so it was necessary to be alert to cues of waning concentration. Interviews were therefore often shorter than perhaps would be anticipated in qualitative interviews generally (ranged from 16 minutes to 2 hours). Wandering concentration also meant I needed to work harder to redirect the participant and manage the direction of the interview. This meant that for some participants it took considerably longer to cover all the topics that I wanted to consider. I found that minimising distractions and interruptions could help to maintaining concentration, for example asking the participant to turn off the television or radio, or suggest closing the door. Sometimes this was beyond my control, such as when talking to participants in the residential care homes staff would often come in to check on the resident or offer refreshment.

Slowed thinking posed another challenge when interviewing. Bourne et al. (2006) suggest using closed questions which require a specific response when talking to people with HD. For some participants open questions were not appropriate and they needed to be given options from which to select an answer. For example I would often start an interview with an invitation like 'tell me a bit about yourself'. For some respondents with HD this kind of open question did not work, and they would give me a simple one line answer. I would then have to use a series of more direct questions to establish their family history, age, marital status, work history, and so on.

Some participants needed additional time to respond to questions. I found waiting for the person to respond testing as I am used to conducting interviews in a more conversational way. I would have to assess if the person was going to respond at all or whether it was just taking time to gather their thoughts and express themselves. In joint interviews, although family members were essential in translating and clarifying information they too were sometimes challenged to wait and allow the person with HD to respond. In this situation, when the person with HD had more advanced

communication difficulties, it required restraint not to focus the interview on the participating family carer but to maintain the involvement of the person with HD. Getting to know the participants over time was an essential part of the study as such built rapport was key to overcoming communication issues (Wilson et al. 2011).

RIGOUR AND GENERALISATION

The rigour of this study is strengthened by the use of triangulation in both the form of multiple methods of data collection (interviews, observations, eco-maps, genograms) and multiple sources of data generation (people with HD, their family caregivers, health and social care professionals). Repeated contact with participants for interviews, observations and eco-mapping supports the rigour of the findings as it has been possible to question participants on more than one occasion. These opportunities for re-interrogation allowed for opinion and events to be reiterated, reinforced, reviewed and checked over time. Participants with HD and their family caregivers were also able to validate their own eco-maps. This form of 'member checking' took place at each interview contact in order to verify and update the eco-maps as care needs, social interactions, and activities shifted over time. This was a multiple case study design and although each case is of interest in itself the aim was to explore a collection of cases so that they might tell us something about the care needs of those affected by HD in the wider context (Stake 1995). Data in this study have therefore been interrogated within and across cases, in several instances, over several years, and throughout data collection.

In order to be able to generalise or replicate the knowledge from this study I needed to provide the reader with sufficient details of the setting and participants (see case profiles in Chapter Five), provide such information in a way that it can be understood and illustrate it with natural experiences from real people (Stake 2000). In this way the reader can recognise transferable

concepts for different settings or communities. There is much discussion about the potential to generalise from case study research (Gomm et al. 2006, Bergen and While 2000, Simons 2009, Thomas 2011). Concepts such as transferability (Lincoln and Guba 2000), naturalistic generalisation (Stake 1995) or replication logic (Yin 2003) are often considered more appropriate for case study research. These concepts support generalisation to theory rather than to empirical data or populations (Sharp 1998). It is this rigour within the data collection and analysis that allows the findings to subsequently be transferred to wider theory. In the findings chapters and discussion I assess how the cases fit with, or develop, proposed theories.

INTERPRETATION

The nature of this study yielded vast amounts of data and I have been necessarily selective in my presentation of those data in order to tell a cohesive story throughout. Not only are the data co-constructed but it should be noted that I alone conducted the analysis and am aware that my own expectations and interpretations (or mis-interpretations) are inevitably part of the reconstructed stories told in this thesis. I have used my own voice in the narrative throughout and I must recognise therefore that it is *my* voice constructing the stories based on the various elements *I* felt to be relevant and important. Hence, these cannot be *participants'* stories about themselves but rather *my* stories about the participants (Rubin and Rubin 2005).

The next chapter presents a brief case profile for each of the 15 cases involved in the study. These profiles provide context for the individual cases and aim to serve as a link between the demographic data presented in this chapter and the cross case synthesis of the qualitative data presented in Chapters Six, Seven and Eight.

CHAPTER FIVE: CASE PROFILES

INTRODUCTION

This chapter provides the case profiles for each of the 15 cases. These summaries include length of time in the study, number and type of interactions, members of the case, genetic family histories and changes over the study period. A genogram was drawn for each case. A key to the symbols used can be found in Figure 9. Where possible the ages of family members have been included for clarification. It was only possible to draw the genograms based on the information given by the participants. Not all could remember all their family ties, and unfortunately there was no way to check this missing data. The focus is on the participants' immediate family for the purposes of identifying previous and potential lines of inheritance of the condition.

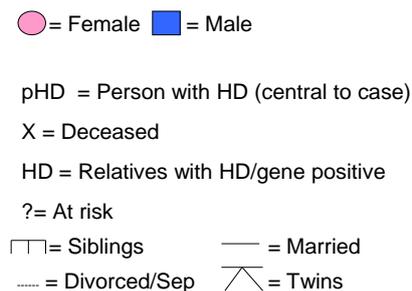


Figure 9 - Genogram symbols

Eco-mapping was used as one of the ways of tracking changes over time. Lines were used to link the central participant (in this case the person with HD) to the other identified people (e.g. main family caregiver, health professionals). The thicker the line the stronger the relationship (1 to 5pt width). A dotted line depicts a particularly weak relationship and a jagged line identifies tension in the relationship (see Figure 10).

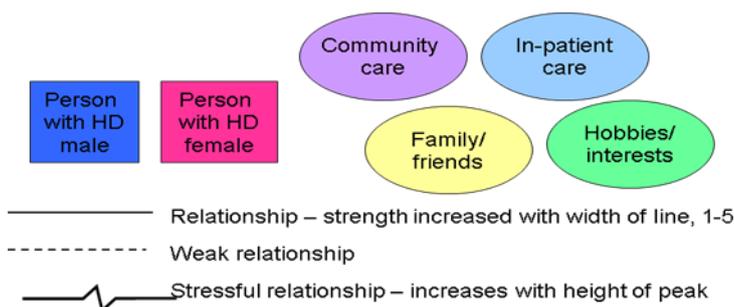


Figure 10 - Eco-map symbols

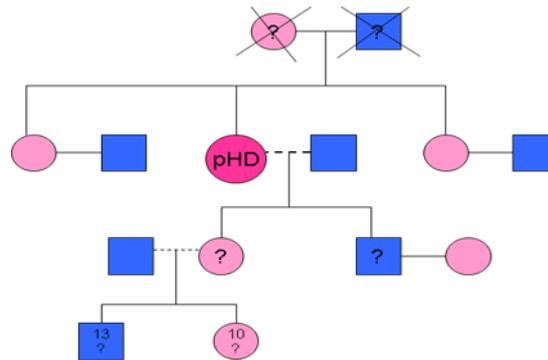
ALISON

Site	Larch Tree Care Homes
Age at diagnosis	54 years
Age at start of study	60 years
Data contribution	19.10.2007 – 19.07.2010
Time in study	2 years 9 months
Number of interviews	5 (individual)
Number of observations	0
Family member interviews	n/a
Health professional interviews	2-support worker at the care centre

Family status: separated, two children, two grandchildren, one older and one younger sister.

Family HD knowledge: Not known: Both parents lived to an old age (mother 95 and father 83)

Genogram



Living situation: Lived at the care centre for two and a half years until it closed. She was relocated in January 2010 to another local care centre run by a different organisation.

Care input: 24 hour nursing care was available at the care centre, with GP support and additional services available.

Key information: Alison moved to the care home when her daughter could no longer manage to care for her at home alongside her two small children. Alison struggled initially to settle into the care centre and missed her family greatly. Distance meant they were unable to visit as often as she would have liked. Alison had several bad falls when living at home so she recognised she needed more care. I met her in October 2007, at that time she had been living at the centre for approximately three months.

Changes over time: Alison had a slow deterioration in abilities and subsequent increase in care needs. In particular Alison's speech, swallow and ability to walk gradually deteriorated over the years she participated in the study. Although level of personal care gradually increased over the study period little significant change was noted. Her family have remained of constant importance to her and continued to visit as regularly as they could. They also took her on short outings and she was still able to leave the care centre to visit them at Christmas. At the new care home Alison no longer had a television and spent time listening to the radio instead.



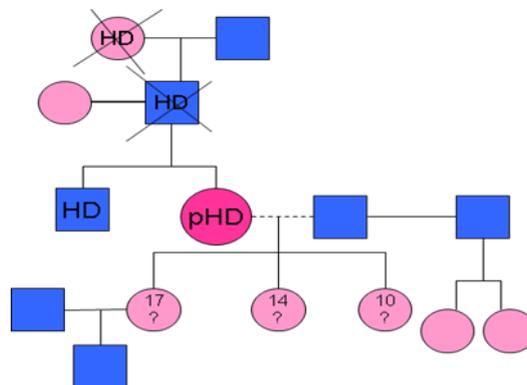
AMY

Site	Oakfield Town
Age at diagnosis	27years
Age at start of study	38 years
Data contribution	31.01.2008-11.02.2011
Time in study	3 years
Number of interviews	3 (joint)
Number of observations	2 clinic appointments, 1 day care visit
Family member interviews	4 husband (3 joint and 1 individual)
Health professional interviews	1 RCA 1 CNS

Family status: Has three daughters from a previous marriage. Her partner, Ben, also has two children from a previous marriage.

Family HD knowledge: Known: Father died with the condition and Amy's younger brother was genetically diagnosed a number of years ago but had not been identified as displaying symptoms.

Genogram



Living situation: Amy lived at home with Ben and her 10 year old daughter Isabelle.

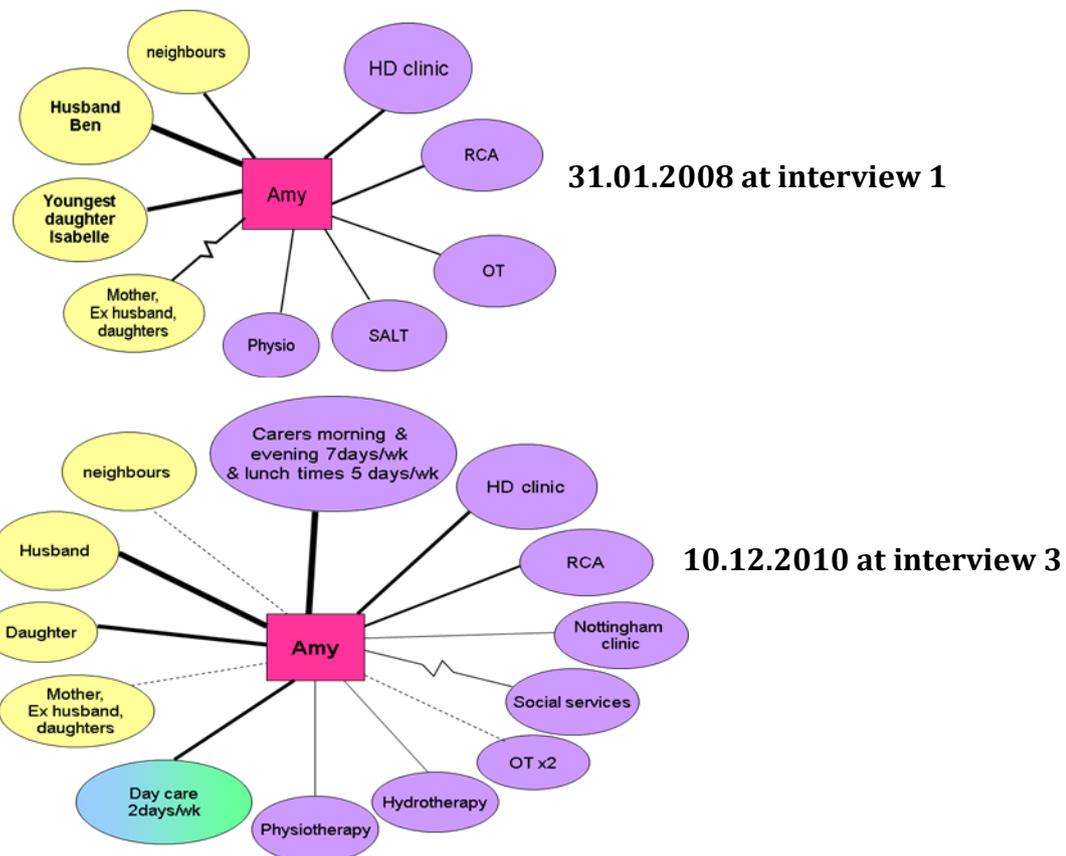
Care input: Amy was cared for by Ben and Isabelle. As her needs increased over the three-year study period a home care package was put in to place to help with washing and dressing in the mornings and evenings. Clinical input was provided by the out-patient clinic at Oakfield Town.

Key information: Amy and Ben had a strained relationship with a number of health and social care professionals during the time they were participating

in the study, and concerns were raised about Isabelle’s welfare throughout this time.

Changes over time: Amy had a number of falls throughout the study period and continually struggled with considerable chorea. She had initially been able to walk most of the time, but by the end of the study was predominately confined to a wheelchair. Her speech also deteriorated considerably. The state provided the family with a disability adapted bungalow in 2008. As the eco-maps show Amy had carers in the mornings, evening and by 2010 also at lunch times. Amy had consistently refused respite care and it was only in the last year of the study that she agreed to attend day care services. As the eco-maps show Amy was accessing a number of services throughout the study period, a notable addition was the hydrotherapy provided at Oakfield Town hospital.

Initially Amy’s relationships with her mother, ex-husband and eldest daughters were strained but this seemed to calm over time, partially as contact with them lessened. Ben was finding caring for Amy increasingly difficult and there was considerable impact for Isabelle, who was often not attending school.



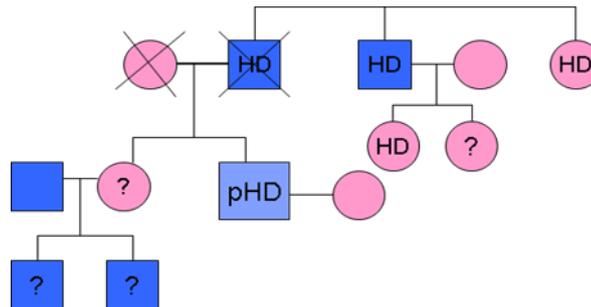
DAVID

Site	Larch Tree Care Homes
Age at diagnosis	41 years
Age at start of study	56 years
Data contribution	02.04.2008-01.06.2011
Time in study	3 years 2 months
Number of interviews	3 (joint)
Number of observations	1 activities session 1 meeting with care providers
Family member interviews	4 wife (3 joint, 1 individual)
Health professional interviews	n/a

Family status: Married to Jill, no children

Family HD knowledge: Unknown: Father died prematurely in an accident but was thought have the condition. Further investigation by David had since led to one aunt, one uncle and one cousin being diagnosed with HD.

Genogram

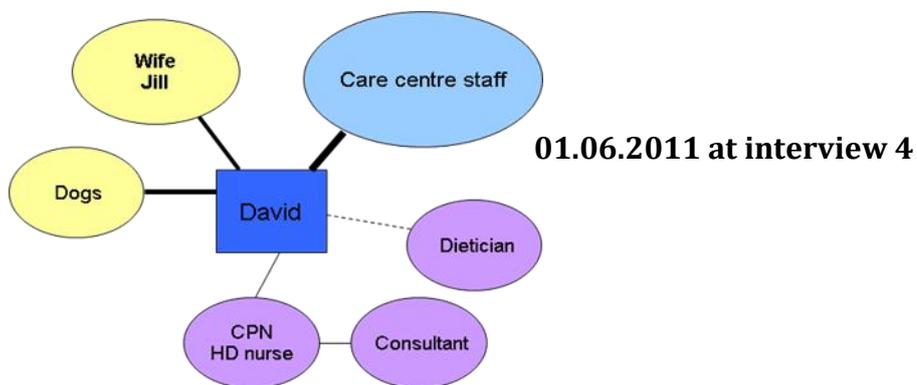
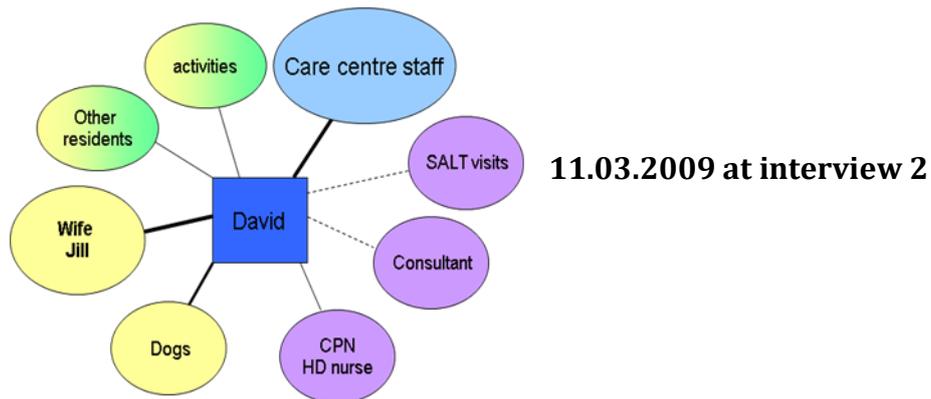


Living situation: David lived at the care centre for the duration of the study.

Care input: 24hr nursing care is available at the care centre. David's care needs gradually increased over the study period, David became increasingly unsteady on his feet, required full help with washing and dressing and had a PEG fitted.

Key information: David moved to the care centre after his wife Jill, could no longer manage his care needs at home. He settled in well and became involved in a number of activities at the centre. Jill trained their two dogs to be Pets as Therapy dogs and they were beloved by a number of the residents. David's condition gradually worsened over time. His wife continued to visit him twice a week and their local CNS for HD also kept in contact to support the family and the care centre staff.

Changes over time: David's progression had been steady over the study period. His walking, speech, swallow and interest in interaction all deteriorated. As the eco-maps show David had input from a speech and language therapist to help maintain his speech and swallow and had ongoing contact with his CNS for HD from his local area, who he had known for a number of years. In the first year of the study period he was still able to attend an out-patient clinic to see a consultant neurologist and regularly went home for the weekend or away for holidays. However, as his final eco-map shows David's condition deteriorated so that the care centre staff increased their input for his physical care. A dietician was involved to monitor his PEG feeds and contact was maintained with his CNS who visited him at the care centre and was able to liaise with the consultant. As David became increasingly disabled it was no longer possible for him to have visits home.



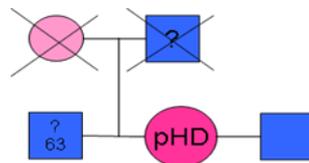
ELIZABETH

Site	Oakfield Town
Age at diagnosis	59 years
Age at start of study	59 years
Data contribution	30.04.2009- 21.07. 2011
Time in study	2 years 3 months
Number of interviews	3 (joint)
Number of observations	3 clinic appointments
Family member interviews	3 husband (joint)
Health professional interviews	2 RCA

Family status: Married to James, no children (Although Elizabeth never mentioned it I was informed that she had given up a daughter for adoption in the 1970s, some efforts were being made to trace her and the couple were discussing the implications with the genetics team).

Family HD knowledge: Not known: Elizabeth's older brother had not been tested but showed no signs of HD aged 63. It is not clear which of her parents had the condition.

Genogram



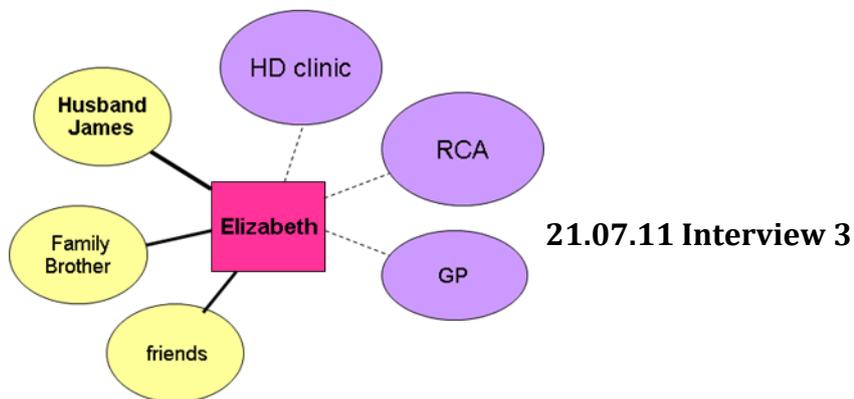
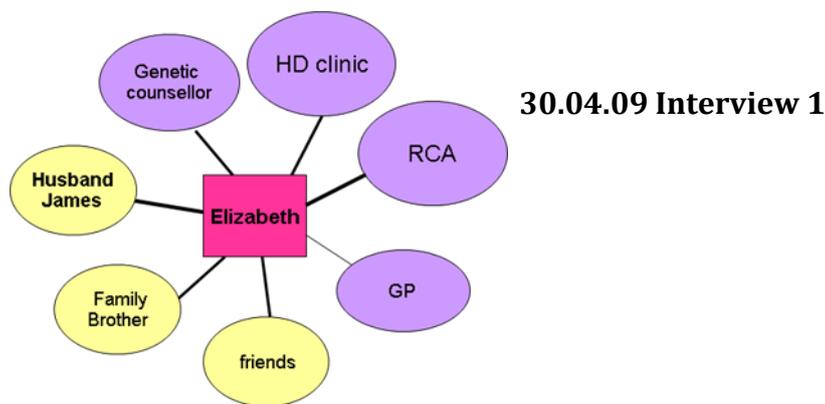
Living situation: Elizabeth lived with her husband in a village in a rural area outside Oakfield Town

Care input: Elizabeth required no care input through the study

Key information: Elizabeth showed few signs of HD and remained at a similar level through the study. She remained able to conduct the majority of daily living activities and was still driving until the last few months of the study period.

Changes over time: There had been little physical change over time for Elizabeth. However, coming to terms with the diagnosis was difficult for both her and her husband, particularly after being told via letter. Their acceptance of this was the main change over time for this couple. It was evident during observations and interviews that they have adjusted well and are keen to

make the most of their life while Elizabeth's condition remained relatively stable. Elizabeth reported that retiring from her job had relieved a number of pressures and improved her mental health. The eco-maps show initial support sought from the genetic counsellor and the RCA reduced over time and links to the clinic remained consistent. In consultation with the couple the consultant reduced their clinic visits from every six months to annually. This occurred approximately eighteen months into the study period. Support from friends and family remained consistent throughout the study period and contact with her GP was minimal.



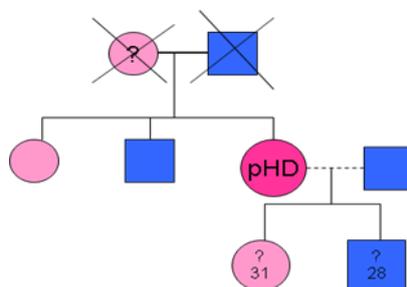
ERICA

Site	Oakfield Town
Age at diagnosis	52 years
Age at start of study	55 years
Data contribution	14.03.2008-20.04.2011
Time in study	3 years 1 month
Number of interviews	4 (individual)
Number of observations	2 clinic appointments 1 emergency clinic visit
Family member interviews	n/a
Health professional interviews	2 RCA

Family status: Divorced with two adult children

Family HD knowledge: Not known: It was thought that the condition traced from Erica's mother's side of the family but this was unconfirmed as her mother had died of cancer when Erica was 16. Erica's father lived until his 70s without showing any signs. Both Erica's brother and sister were tested and do not have the Huntington's gene. However both Erica's children are at risk but had chosen not to be tested.

Genogram

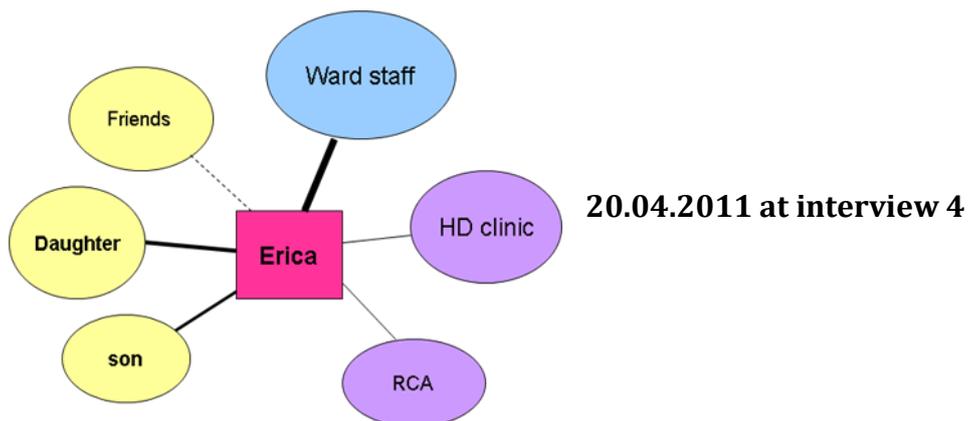
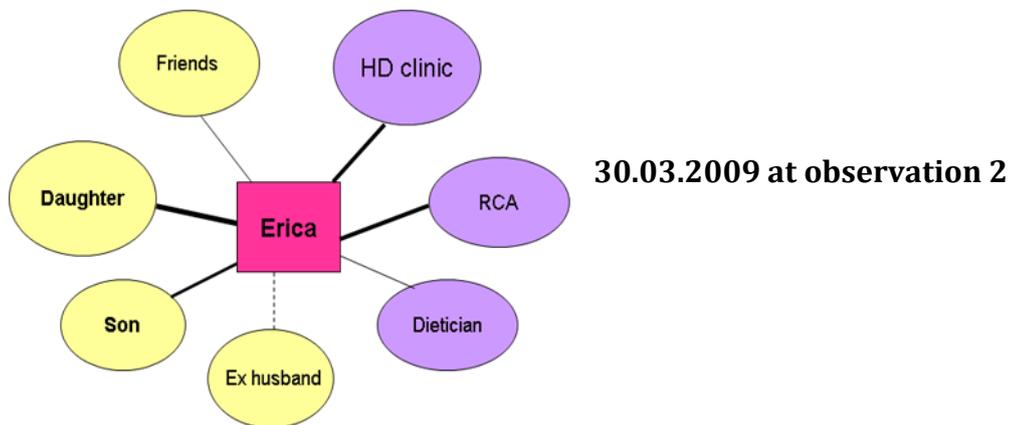


Living situation: Erica lived alone, although throughout the study period her daughter was living with her for a short time.

Care input: Erica lived independently at home until the end of the study period when she had a serious psychiatric episode and required emergency hospital treatment. She remained at the psychiatric unit at the hospital beyond the end of her involvement in the study.

Key information: The key incident for Erica was her psychiatric episode in January 2011. Until this time she had appeared to be managing her condition well.

Changes over time: Generally Erica’s health had been very stable over the three-year study period. As the eco-maps show, Erica lived at home with some supportive input from the local HD services and periodically a dietician. She had support from her daughter and son but required no help at home. In January 2011 Erica was brought to clinic for an emergency appointment by her two children, suffering from a severe psychiatric episode, she reported hearing voices and was completely unable to manage her personal care or activities of daily living. She was admitted immediately and was cared for in a psychiatric ward at the hospital, with 24 hour support from staff. She also required some help with activities of daily living and was not considered to be able to look after herself at home. Her children remained involved and visited regularly.



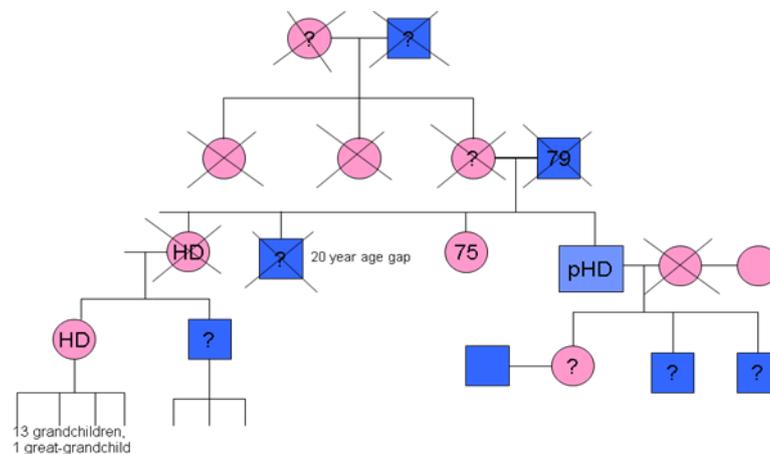
HARRY

Site	Birchtree City
Age at diagnosis	69 years
Age at start of study	72 years
Data contribution	17.10.2007 – 30.09.2010
Time in study	3 years
Number of interviews	3 (joint)
Number of observations	4 CNS home visits 3 Clinic appointments
Family member interviews	3 wife (joint)
Health professional interviews	1-CNS

Family status: Widowed and re-married to Margaret. Three adult children from his first marriage.

Family HD knowledge: Not known: Harry's children have not been tested for the HD gene. Some investigation revealed Harry's eldest sister (there was a 20 years age gap between them) did have HD, as does her daughter. It is suspected that the HD came from his mother's side of the family but this is unconfirmed.

Genogram



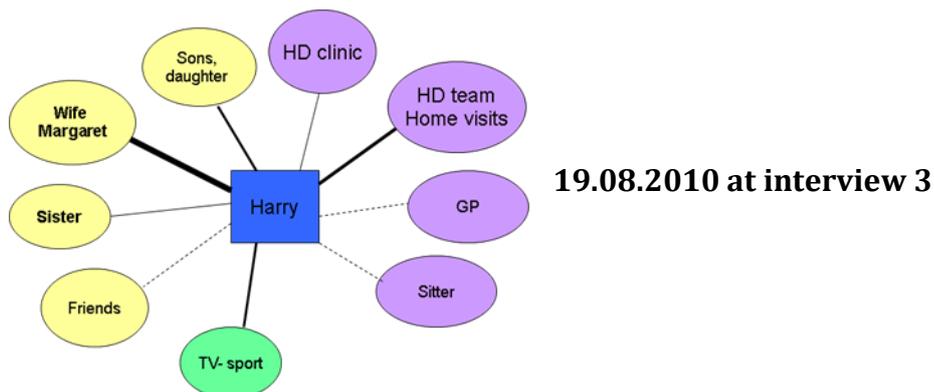
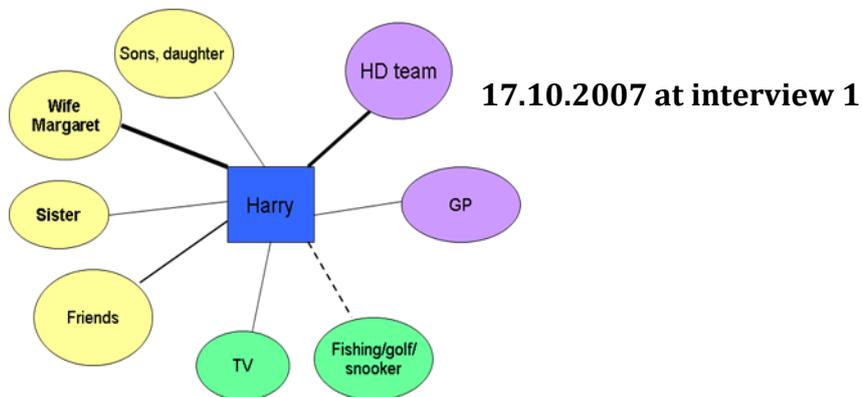
Living situation: Harry lives with his wife, Margaret, in their house in a suburb of the city.

Care input: Margaret has been his main carer throughout the study. The couple used a sitter service for a short period but they found inflexible visiting times hindered their use of the service. They had input from the CNS who visited them at home. Harry also had yearly out-patient clinic appointments with the Birchtree team and access to additional input from

other services, such as speech and language therapy and dietetics, as and when necessary.

Key information: Harry was an older man who was contending with the normal deficits of old age alongside his HD. Margaret was also in her 70s and suffered from a number of health complaints, including a heart condition. Harry required no help with personal care. However, his memory became progressively worse so that he could not be left alone for long periods.

Changes over time: Harry slowly lost interest in the things he enjoyed and lacked motivation to go and do things. As the eco-maps show he reduced and finally stopped participating in his hobbies. Harry went through a period when he became extremely anxious when he was due to go out to places where he would not know people. Consultation at clinic and some medication mostly resolved this issue. One aspect was to increase the involvement of Harry's three adult children to allow Margaret to take breaks. The eco-map also shows the relationship with his sons and daughter becoming stronger over time. Harry's dependence on Margaret also increased over time as he needed more support with taking medication, preparing meals and the tasks of daily living.



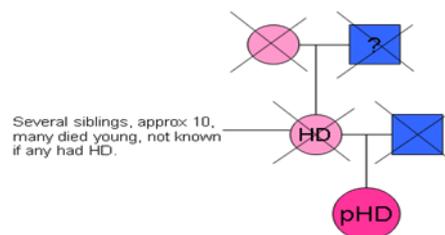
HELEN

Site	Birchtree City
Age at diagnosis	68 years
Age at start of study	70 years
Data contribution	11.12.07 – 13.12.10
Time in study	3 years
Number of interviews	4 (individual)
Number of observations	3 clinic appointments
Family member interviews	n/a
Health professional interviews	2-CNS

Family status: Single, no children or siblings

Family HD knowledge: Known: Helen’s mother died with HD. Helen’s mother had a number of siblings, many of whom died young. Helen was not able to pin point exactly how many and approximated this at about 10.

Genogram

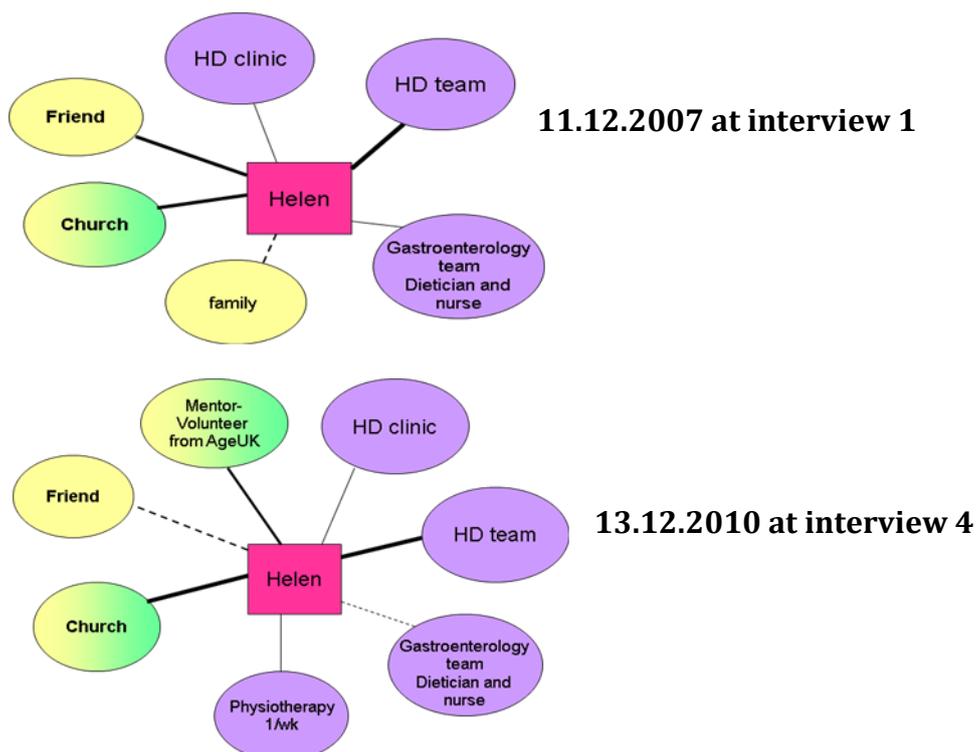


Living situation: Helen lived alone in her own home, just outside of the city centre.

Care input: Helen did not require any care input throughout the study period although had support and advice from the CNS within the Birchtree HD team.

Key information: Helen was an older woman living alone and was newly diagnosed when she started in the study. She had a known family history of HD and cared for her mother until she died in 1980. The CNS had been involved in supporting her to manage relationships with friends and members of her church since her diagnosis. She has also encouraged Helen to make some minor adjustments in order for her to remain in her home for as long as possible. The fact that she lived alone was of concern to the CNS and she preferred to maintain close contact with Helen to make sure she was managing activities of daily living.

Changes over time: Helen showed slow deterioration over the study period. She became unsteady on her feet but was still able to get out and about and manage the majority of tasks of daily living. During this time she took someone on to do her ironing and had meals delivered to help with food preparation. Helen noted some deterioration in her concentration and memory. During the study period Helen’s driving license was revoked and this restricted her activities. However she still made an effort to get out by using taxis and a network of friends for lifts. She continued to enjoy walking but did not felt confident to do this alone. The introduction of a volunteer from AgeUK was very successful and Helen was excited for this to continue so that they could go walking together. This limited change in Helen’s care input and activities is reflected in the eco-maps. One area of note is the change in her relationship with an important friend. This previously strong relationship became quite fractious and resulted in Helen no longer spending much time with this person. Helen’s relationship with people at church, although difficult at times remained strong and this was a key part of her life. The HD team and yearly clinic visits also remained stable and were her main source of professional input. An addition to this was the introduction of weekly physiotherapy sessions in her home in order to help her maintain core strength for walking and balance. Helen also maintained links with the gastroenterology team for an existing condition which restricts her diet.



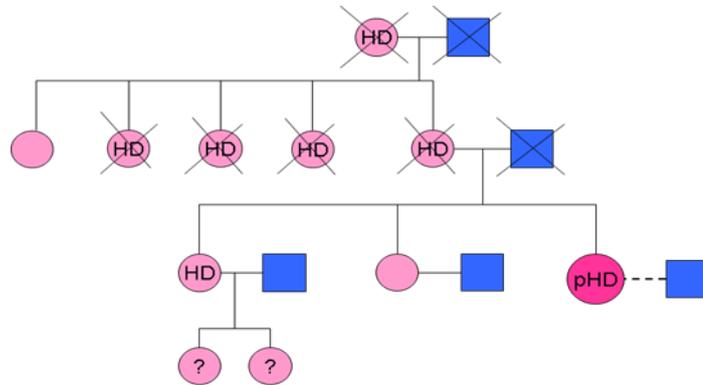
KATE

Site	Larch Tree Care Homes
Age at diagnosis	30 years
Age at start of study	42 years
Data contribution	14.06.2007 - 20.07.2010
Time in study	3 years
Number of interviews	4 (individual)
Number of observations	1 arts and crafts session, 1 tea party
Family member interviews	n/a
Health professional interviews	2-staff nurse at the care centre

Family status: Divorced, no children, has two older sisters

Family HD knowledge: Known: Mother and several aunts died with HD, one sister had also been diagnosed

Genogram



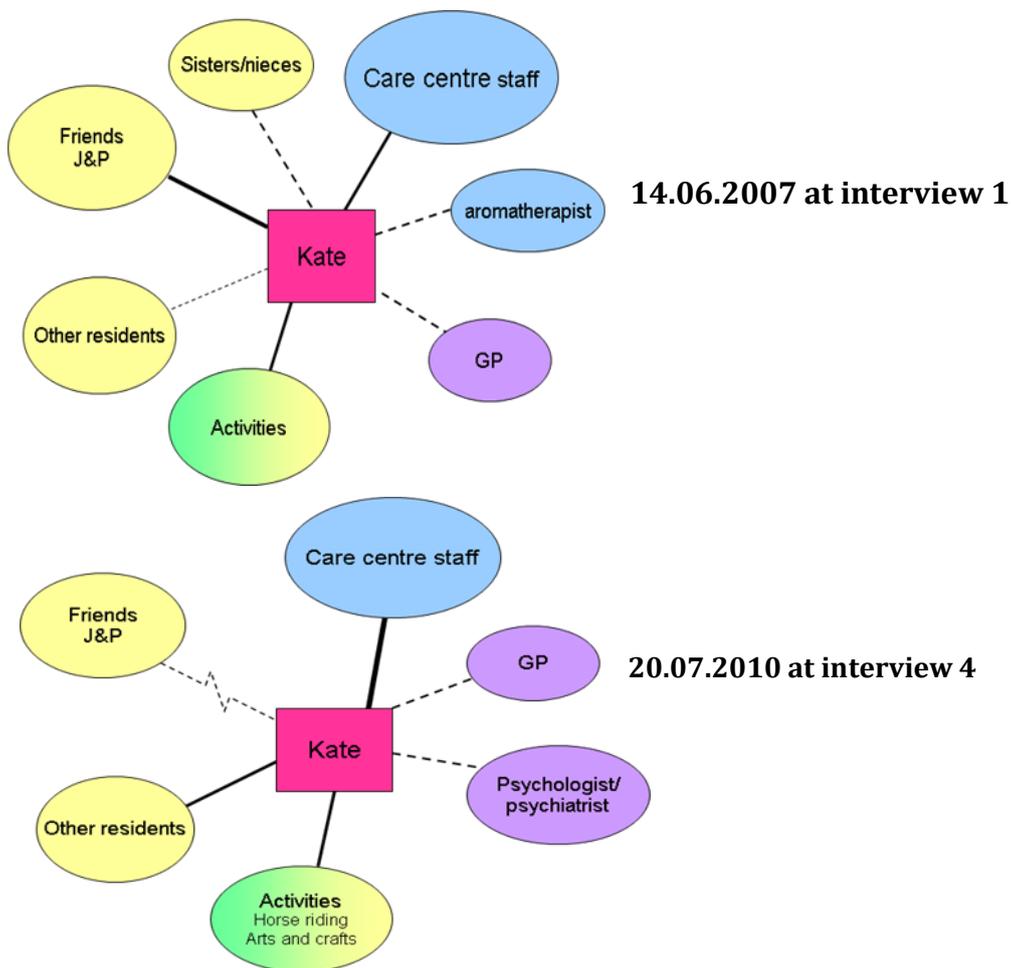
Living situation: Kate moved to the care centre in 2003. After struggling to cope on her own, she made the decision to find somewhere she could have 24 hour care.

Care input: 24 hour nursing care was available at care centre with GP support and additional services when required.

Key information: Kate was physically quite well but presented with more cognitive and behavioural issues. She had limited contact with her family and she felt this was because they were unable to cope with the family diagnosis.

Changes over time: As the two eco-maps show, the care centre staff were central to Kate's care. The increase in the thickness of the line linking this group shows their increased involvement in Kate's personal care as her needs progressed. Family have had little contact during the study period and by the end were not involved at all. Relations with two key friends who had

been an important part of Kate’s social support became increasing difficult over the study period and by the time of the final interview they had no contact. Activities and other residents were important and her involvement with them remained consistent over the study period. GP input was provided to the care centre, but this remained a peripheral service throughout. A notable change was the addition of psychiatric services in her final year in the study. The care home requested this psychiatric input as they were struggling to manage Kate’s behaviour.



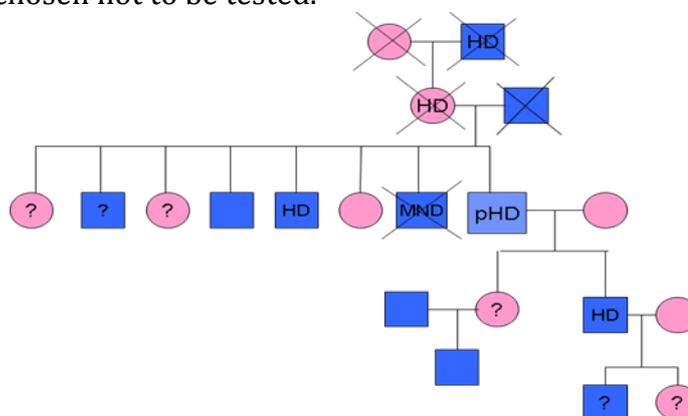
MARTIN

Site	Birchtree City
Age at diagnosis	55 years
Age at start of study	60 years
Data contribution	12.02.2008-19.04.2011
Time in study	3 years 2 months
Number of interviews	3 (joint)
Number of observations	1 CNS home visit 1 Clinic appointments 2 SALT home visits
Family member interviews	3 wife (3 joint, 1 individual)
Health professional interviews	2 CNS 1 SALT

Family status: Married to Julia, two adult children and two grandchildren with a third born during the study period.

Family HD knowledge: Known. Martin's mother and grandfather died with condition and at least one of Martin seven siblings has been diagnosed. Martin and Julia's son was genetically but not clinically diagnosed. Their daughter had chosen not to be tested.

Genogram



Living situation: Martin lived at home with his wife in a suburb of Birchtree.

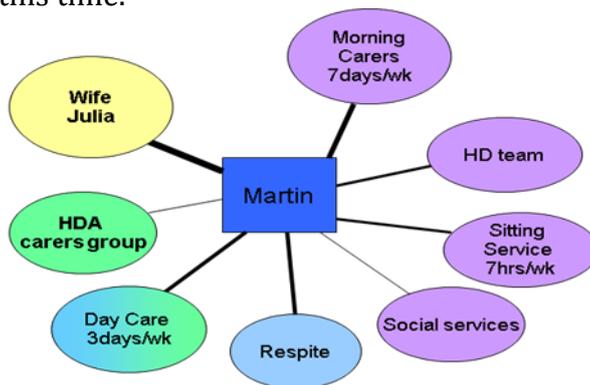
Care input: Martin was cared for at home by his wife with additional care input from a number of services. He initially had carers come in the morning to help with washing and dressings, as his condition progressed overnight carers were also added. Martin also accessed a day care centre three times a week, had several weeks respite allocated per year and used a sitting service seven hours a week as he could not be left on his own. He also had input from

physiotherapy, Social Services, the Birchtree HD team and increasingly from the speech and language therapist.

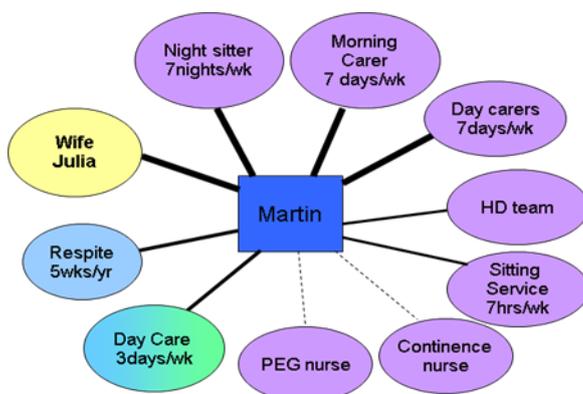
Key information: Martin’s wife was his main carer and had a number of serious health issues of her own including a heart condition and diabetes. Towards the end of the study he was struggling to swallow and had a PEG fitted. However he suffered from a number of chest infections and ultimately died in hospital shortly after his completion in the study.

Changes over time: Martin’s condition clearly worsened over the study period. Initially he had been walking, eating well and communicating. However all of these functions deteriorated. He required a wheelchair and specialist bed. Overnight carers were put in place as he also suffered with disturbed sleep and could not be left alone at night. Martin’s speech and cognition also declined over the study period and he was able to make little contribution to interviews towards the end of the three years.

Martin’s final eco-map shows substantial care input from morning, night and day carers. There was a discernable shift from services such as physiotherapy and speech and language therapy to the input from the continence nurse and the PEG nursing team. The eco-maps also show the number of community services needed in order to allow Martin to continue to live at home as his condition progressed. The Birchtree HD team remained involved throughout this time.



12.02.2008 at interview 1



20.07.2010 at interview 4

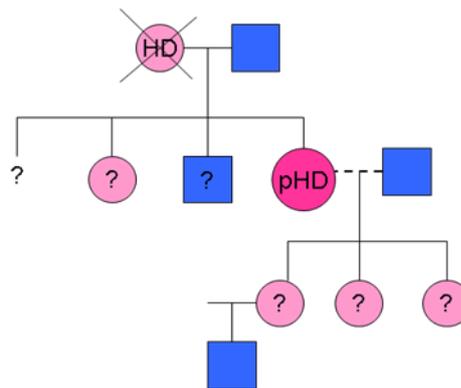
MARY

Site	Larch Tree Care Homes
Age at diagnosis	40 years
Age at start of study	51 years
Data contribution	11.02.2008 – 01.2009
Time in study	11 months
Number of interviews	1 (individual)
Number of observations	1 physiotherapy session
Family member interviews	n/a
Health professional interviews	1 staff nurse at the care centre

Family status: Divorced, three siblings, three daughters and a grandson and one granddaughter (born during the study period)

Family HD knowledge: Known: Mary's mother died with the condition, her siblings and daughters had not been tested.

Genogram

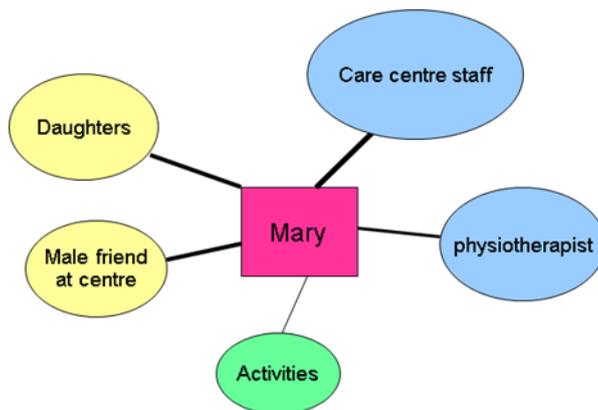


Living situation: Mary came into the care centre for respite in December 2007, however it became clear that she would not have been able to go home as she lived alone and could not have managed by herself. Mary remained at the care centre for approximately 13 months however after displaying a number of behavioural problems staff felt they could no longer manage her behaviour at the centre and a place was found for her at a psychiatric unit nearer her family. Although she had expressed a wish to move to the unit Mary was ultimately sectioned under the Mental Health Act and transferred to the unit in January 2009.

Care input: 24 hour nursing care was provided at the care centre with GP support. Personal care was required and additional psychiatric services were called in response to challenging behaviour.

Key information: Mary did not wish to be at the care centre and often made attempts to leave. Her behaviour was particularly challenging for staff to manage.

Changes over time: Mary was only at the care centre for a short time and never fully consented to being there. During her time at the care centre Mary became increasingly unsteady on her feet and attended physiotherapy sessions to maintain her strength and help her co-ordinate the use of a wheelchair. Through her physiotherapy she met and became close with a male resident.



01.08.2008 at observation 1

As Mary was only in the study for a short period she does not have comparable eco-maps. The one above was collated during an observation session and depicts Mary's reliance on the care centre staff for physical care. The eco-map also shows how much she valued the support of her daughters and a male friend at the centre. She rated physiotherapy as central to her care and had a good relationship with the physiotherapist. Mary also participated in some of the care centres provided activities.

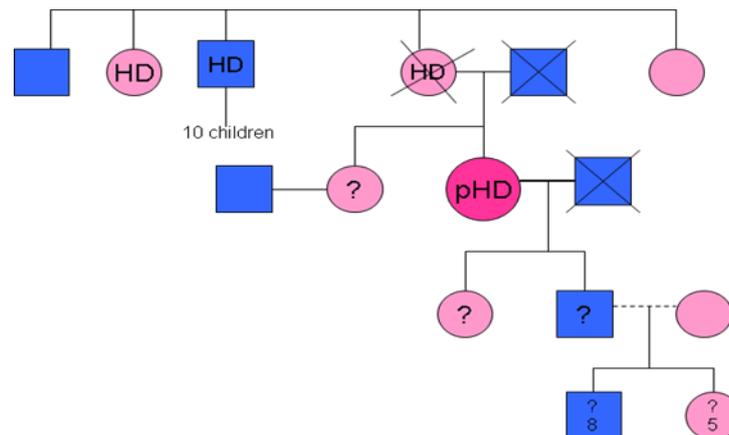
ROSE

Site	Birchtree City
Age at diagnosis	60 years
Age at start of study	61 years
Data contribution	22.02.2008- 09.03.2011
Time in study	3 years
Number of interviews	1 (1 individual, 2 joint)
Number of observations	1 CNS home visit 3 clinic appointments
Family member interviews	2 daughter (2 joint)
Health professional interviews	1 CNS 1 rehabilitation support worker (RSW)

Family status: Widow, with two adult children.

Family HD knowledge: Known: Rose's mother died with the disease, one uncle and one aunt were thought to have the condition. Neither of her children had been tested.

Genogram



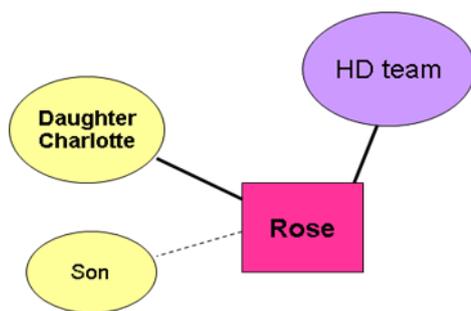
Living situation: Rose lived with her adult daughter, Charlotte in their home.

Care input: Rose did not require any physical care input throughout the study period. She was able to carry out most tasks of daily living but her daughter provided input in the form of cooking and house work. Rose attended the out-patient clinic on a yearly basis and had home visits from the CNS or rehabilitation support worker approximately every 6 weeks.

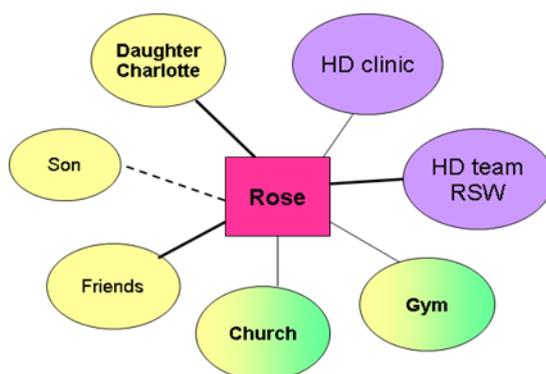
Key information: Rose was physically quite able but suffered from a form of vertigo, the cause of which was unknown. The HD team did not think it was a physical symptom of the condition however other specialities were unable to

find a cause or solution to this issue, and finally concluded that it was part of her HD. Rose was still able to drive and maintained some contact with her social circle, although she felt the vertigo restricted her life.

Changes over time: Rose showed very little change over time. The vertigo was continually an issue for her and her daughter because of the restrictions it placed on her mobility. Input from the rehabilitation support worker encouraged her to use a gym and walker to improve her mobility. As can be seen in Rose's eco-maps the focus of her care input was on encouraging and supporting her to regain her social life and activities. The rehabilitation support worker took a key role in this by establishing ways in which she could regain her confidence to do the things she wanted while maintaining her safety and contributions within the home and community.



22.02.2008 at interview 1



09.03.2011 at interview 3

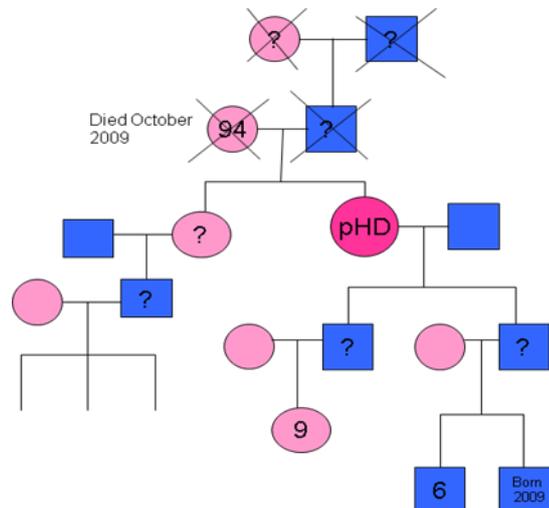
SARAH

Site	Birchtree City
Age at diagnosis	55 years
Age at start of study	61 years
Data contribution	27.11.07-04.11.10
Time in study	3 years
Number of interviews	2 (joint)
Number of observations	1 home care visit, 3 clinic appointments
Family member interviews	5 husband (2 joint, 3 individual)
Health professional interviews	1 CNS 1 staff nurse at the care centre

Family status: Married to Max, has two adult sons and three grandchildren (one born during the study period)

Family HD knowledge: Not known: Mother died at the age of 94 (during the study period), father died age 71 so it was suspected to be from his side of the family. Neither son has been tested.

Genogram



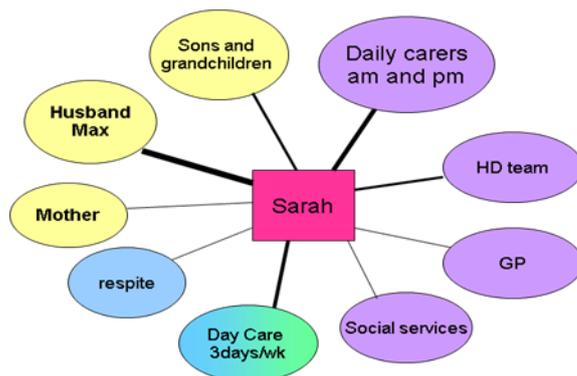
Living situation: Sarah lived at home with her husband until the end of 2007 when Max suffered a stroke and could no longer care for Sarah at home. She moved to a residential care centre in the local area and remained there for the rest of the study period.

Care input: At home Sarah was predominately cared for by her husband and had a package of care including morning and evening carers to help her to get up and washed and to help her to bed. She also attended day care three days

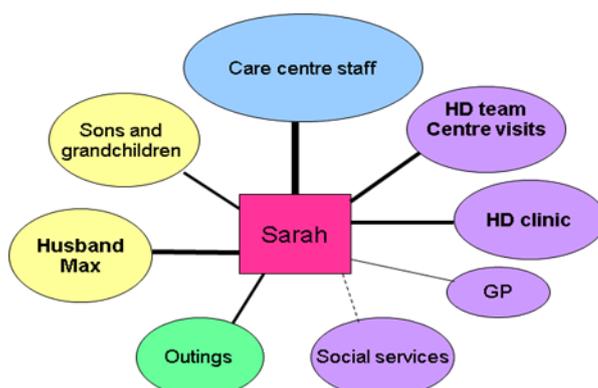
a week and had access to six weeks respite per year. After moving to residential care 24 hour nursing care and support was provided by the care centre staff. She continued to have contact with the HD team, being visited at the centre by the CNS and dietician and attending out-patient clinics for medication reviews as and when necessary.

Key information: The key issue for Sarah was her transition to the care centre as this had an impact for both her and her husband.

Changes over time: Over the study period Sarah showed steady deterioration. She became increasingly unstable on her feet and began to use her wheelchair more. Her mental capacity also deteriorated and it became increasingly difficult for her to participate in study interviews. Sarah had several problems with eating and this was assessed to be a behavioural issue rather than physical deterioration. The first eco-map was compiled with Sarah's input while she was still living at home. At this point she had a number of community services for her needs and relied on her husband for the majority of her care. Moving to the care centre meant that the care centre staff then took a predominate role in her daily care but community services remained involved. Socially Sarah always enjoyed outings provided by the care centre and continued to have a high level of input from her family, with her husband visiting at least twice a week.



27.11.2007 at interview 1



16.07.2010 at interview 4 (husband only)

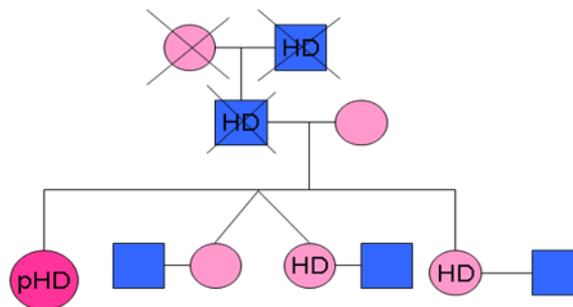
SOPHIE

Site	Birchtree City
Age at diagnosis	40 years
Age at start of study	46 years
Data contribution	16.06.2008-02.06.2011
Time in study	3 years
Number of interviews	2 (joint)
Number of observations	2 clinic appointments 1 SALT home visit
Family member interviews	4 sister (2 joint, 2 individual)
Health professional interviews	1 CNS

Family status: Single, no children.

Family HD knowledge: Known: Father died with the condition, two of her three sisters have the condition.

Genogram



Living situation: Sophie lived with her sister, Melanie, and her brother-in-law.

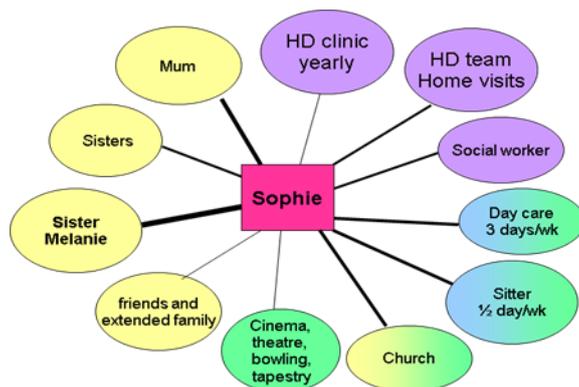
Care input: For the majority of the study period Sophie required little care input. She was able to carry out personal care and managed some activities of daily living with assistance from her sister. As her condition deteriorated she had increased input from occupational therapy, which assessed the kinds of equipment that could be put in place to help Sophie continue to live at home.

Key information: Sophie's condition remained fairly stable throughout the study period. She had input from physiotherapy, a speech and language therapist and a dietician to help maintain as much of her functioning as possible. Sophie was still able to walk but with an unsteady gait, and her speech although impaired, was understandable. She also displayed some cognitive deterioration throughout the period of study. She continued to be

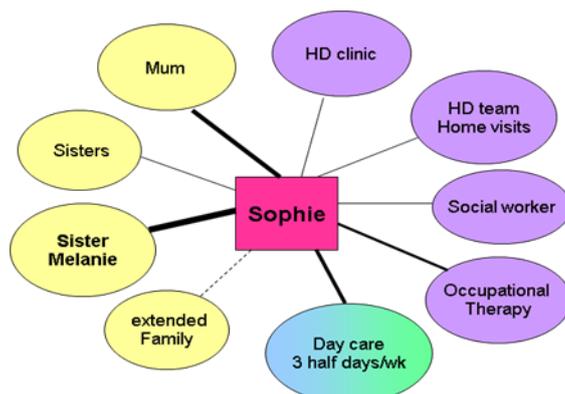
involved in a number of social and respite activities, although these were curtailed towards the end of the study period when funding for them was considerable reduced. Living with her sister caused some tensions, particularly as Melanie was newly married. Sophie's mother and Melanie's twin sister (also suffering with HD) lived nearby, allowing Melanie and her mother to help each other in caring for Sophie and her sister.

Changes over time: Sophie increasingly required help with her activities of daily living and by the end of the study needed help with personal care and could no longer be left on her own even for short periods. She began to have a number of falls and was increasingly dependent on her wheelchair. Changes in funding to both the care home Sophie used for respite and the day care services placed increasing pressure on Melanie. Around this period Melanie was made redundant so it was not possible for the family to fund these services for themselves.

Over time Sophie was less able to participate in the activities which supported her socially and the dependence on her sister for her daily care needs increased. Occupational therapy also became involved in Sophie's care as she required more assistance to live well at home. Towards the end of the study occupational therapy were providing a shower chair and bed rails.



12.03.2009 at interview 2



01.06.2011 at interview 4

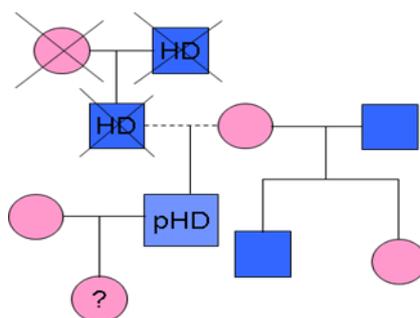
TOM

Site	Birchtree City
Age at diagnosis	45 years
Age at start of study	46 years
Data contribution	21.10.2008-22.06.2011
Time in study	2 years 8 months
Number of interviews	3 (3 joint, wife joined late on 2 occasions)
Number of observations	2 clinic appointments 1 CNS home visit
Family member interviews	3 wife (joint, 1 with daughter as well)
Health professional interviews	2 CNS

Family status: Tom was married with one adult daughter

Family HD knowledge: Unknown: Tom's father committed suicide when Tom was a young child, his mother remarried, contact was not maintained with his father's side of the family.

Genogram

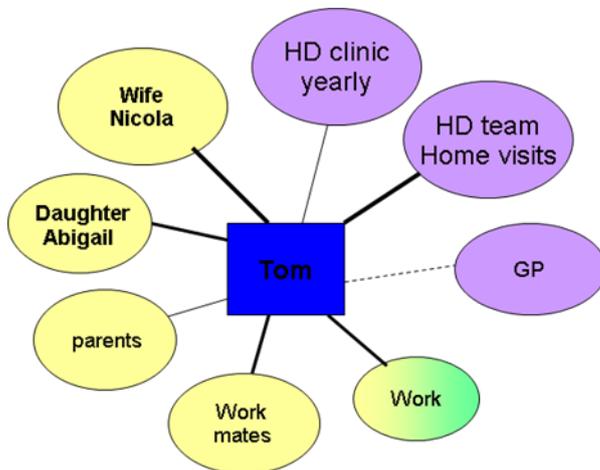


Living situation: Tom lived at home with his wife, Nicola and daughter, Abigail.

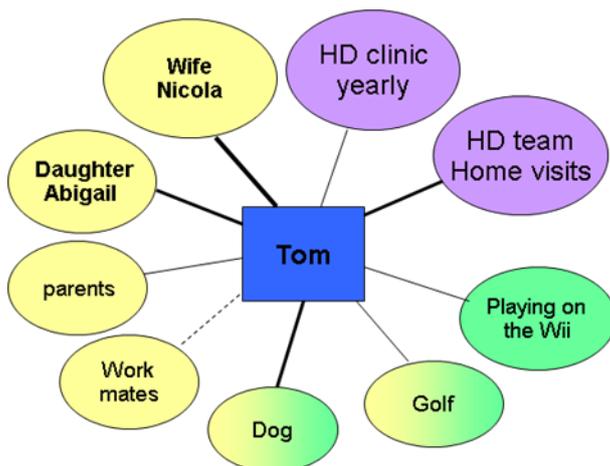
Care input: Tom required no help with personal care and was able to carry out all his activities of daily living. He had some input from the rehabilitation support worker and support from the CNS.

Key information: During the first year of study period Tom was still working. Finishing work was an important transition for Tom and this was supported by the CNS. Her role was also valued for providing the family with information about the condition and helping them to adjust to the changes in Tom's behaviour.

Changes over time: As Tom was newly diagnosed he experienced little physical change over the study period but focus was on him and his family adjusting to living with the condition. The loss of Tom’s job was also a pivotal time for the family. The eco-maps show how Tom needed to find ways to keep active after finishing work. The family got a dog and Tom walked him twice a day. He had always played a number of sports and with the help of the rehabilitation support worker began to play golf on a regular basis as well as a number of games on the Wii in order to maintain his motor skills, core strength and muscle development. Reliance on the HD team reduced over time as Tom and his family came to terms with his condition, became more confident and independent.



21.10.2008 at interview 1



22.06.2011 at interview 3

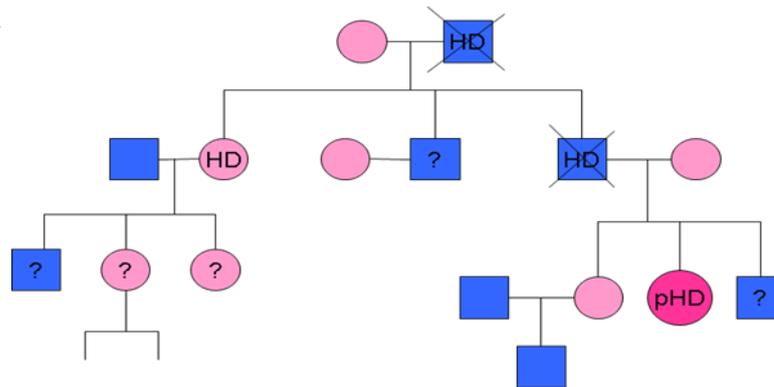
VICKY

Site	Oakfield Town
Age at diagnosis	19 years
Age at start of study	26 years
Data contribution	17.12.2008-14.07.2011
Time in study	2 years 7 months
Number of interviews	2 (individual)
Number of observations	2 clinic appointments
Family member interviews	3 mother (individual)
Health professional interviews	2 RCA

Family status: Vicky was not married and had no children

Family HD knowledge: Known: Vicky's father and grandfather died with the condition, her aunt was also affected. Her older sister had been tested and did not have the affected gene, her younger brother had not been tested.

Genogram



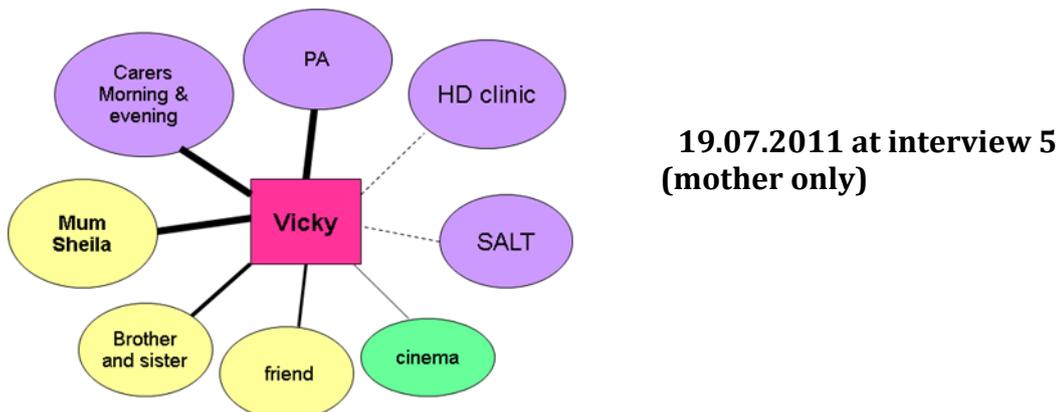
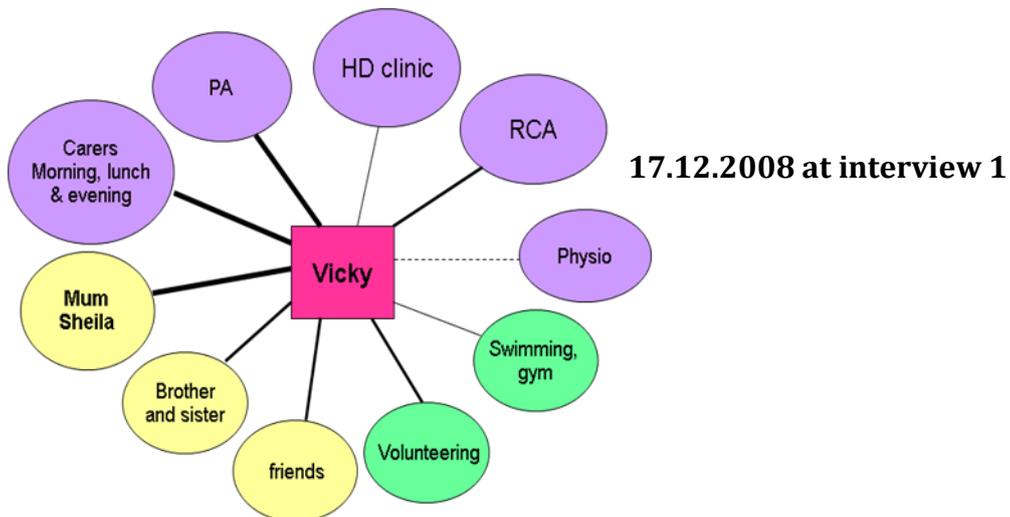
Living situation: Vicky lived alone in an adapted flat.

Care input: Vicky had a personal assistant five days a week to take her out during the day. She also had carers in the morning and evenings to get her up, washed and help prepare meals. Her mother, Sheila was also very involved in her care and would visit a number of times a week.

Key information: Vicky lived alone with a care package in place to tend to her personal care and some activities of daily living, as the study period progressed Vicky's care needs increased. She had initially been quite independent and lived with the support of a personal assistant and morning and evening carers. Over time her speech, gait, swallow, cognition and

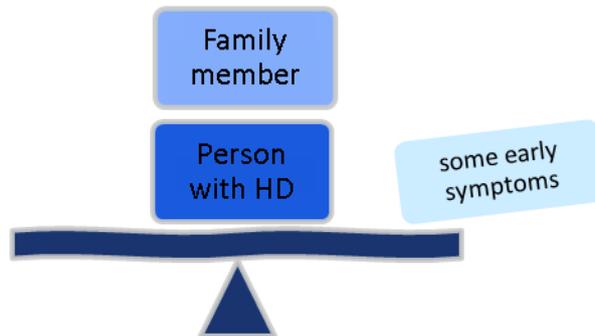
behaviour deteriorated so that it was increasingly difficult for Vicky to remain in her flat alone.

Changes over time: As the eco-maps show, over the period of the study Vicky's reliance on her mum, the carers and her personal assistant increased. As her abilities decreased she was no longer able to participate in some of the social activities she enjoyed, such as the children's holiday club she had volunteered for and physical activities such as swimming and going to the gym. Vicky still enjoyed one-to-one outings with her personal assistant such as going to the cinema. Her involvement with the HD clinic also decreased as Vicky's needs became more immediately hands on. She required increased help with personal care such as getting up, washed, dressed and at meal times. Vicky had no care input at the weekends and this was becoming an increasing concern for her mother. Although living independently throughout the study period it was becoming increasing clear that to continue to do so she would require even more care input.



QUALITATIVE FINDINGS

CHAPTER SIX: DISCOVERY AND ADJUSTMENT



Early phase – balancing discovery and adjustment

INTRODUCTION

The focus of this chapter is on those who were more recently diagnosed and predominantly remained in what can be considered the early phase of the condition (see Table 10). This chapter is constructed to address four elements illustrated by participants in this early phase. Initially the chapter addresses the diagnosis as a dramatic and distressing event, whether there was knowledge of the condition in the family or not. A second section shows how responses to this diagnosis were both emotional and practical and additional contributions are made from family members. The third section looks at the management of this new knowledge. Patients and families can be seen to use strategies of information seeking, controlling, blocking and resisting.

The fourth part of the chapter focuses on input from health and social care professionals during this early phase. The focus here is on how and why professionals build knowledge of the patient and their family, the level of care they provide, the importance of the key worker role and how professionals

balance the needs of patients and their family cares as well as engaging in care planning for future care and support.

Table 10 - Cases to illustrate knowledge management

Name	Age at diagnosis	Time since diagnosis at start of study	Living situation	Care needs
Elizabeth	59	1	Lived with her husband, James	Some help needed with activities involving fine motor skills
Erica	52	3	Lived alone until she had a psychiatric breakdown and was admitted to a unit at the hospital as discussed in Chapter Six	Reported not needing any help with activities of daily living or home management
Harry	66	3	Lived with his elderly wife Margaret	Required some help with preparing meals and encouragement to undertake activities
Helen	68	2	Lived alone	Had help with ironing and meal preparation.
Rose	60	1	Lived with her daughter, Charlotte	Required some help with preparing meals and encouragement to undertake activities
Tom	43	1	Lived with his wife, Nicola and their adult daughter, Abigail	Some help needed with activities involving fine motor skills

THE DIAGNOSIS

All participants had different routes through the health care system to their diagnosis. The majority, like Harry, Erica, Tom and Elizabeth, were referred to a neurological out-patient clinic after presenting to their GPs with symptoms. Others, such as Rose, had prior links to services due to their family experience of HD. Not all participants were able to identify the exact date of their diagnosis but time since ranged from approximately 8 months to 15 years. For a number of participants the process of diagnosis was not simple, and for a few it took months and even years to identify HD as the source of their symptoms. Each of the participants highlighted in this chapter talked about presenting with

symptoms which prompted their eventual diagnosis. People particularly identified clumsiness and reduced fine motor control, twitching movements when at rest and reduced ability to co-ordinate multiple tasks as initial symptoms. These factors finally culminated in people seeking a diagnosis. However these mild initial symptoms often persisted for a number of years during which they were ignored, or put down to other things such as stress, advancing age, or even accepted as 'just how that person is'.

I was clumsy with coffee at work, because I was going through the change [menopause]. ... And I just assumed it was that to do with the [menopause]. (Erica, Interview 1, 14.03.2008)

Well when I first met him ...one of the first things you told me about yourself, was that you were rather clumsy. But of course I didn't think anything of it at the time. We noticed pots getting chipped and things cracked and little things happening, that you didn't really put it down to anybody or anything, but over a period of time, when you look back you realise that it was all part of it. (Margaret, Harry's wife, Interview 1 17.10.2007)

Family members were encouraged to look back to when they felt symptoms started to appear. Some mentioned how the diagnosis had now clarified incidents in the past or identified certain behaviours as part of the condition.

Because I assume now it could well be possibly because of Huntington's, you had a couple of falls, and at work you fractured your knee didn't you? (James, Elizabeth's husband, Interview 1 30.04.2009)

Helen and Harry had their symptoms noticed by other health professionals involved in their care who then alerted them to seek further help. Helen had been visiting an out-patient clinic for another condition when her unsteady gait was recognised, aware of her mother's condition she needed little further confirmation, as she explains in this section;

... [the consultant] said 'would you please walk back to the doorway and walk back in again'. Well of course my heart sank. I knew immediately

what it was. And he'd noticed something different. And of course I'd, rather unusually for me for once in my life I was speechless and I just sort of stopped dead and he just said it again, just repeated. He could tell I wasn't deaf, looking very puzzled by this time, and then I just took a deep breath and said 'my mother had Huntington's disease'. (Helen, Interview 1, 11.12.2007)

RESPONSES TO NEW INFORMATION

SHOCK AND DENIAL

For those aware of their 50% risk of having the HD gene, even if they were aware they had some symptoms of the condition; the official diagnosis was still a tremendous shock. In her first interview Helen explained how despite knowing she had a 50% chance of having the condition and recognising some of the symptoms she was shocked when she received her diagnosis. Three years later in her fourth and final interview she reflects on her reaction.

Helen: I was thinking now why was it a shock to me when I was diagnosed, when I'd obviously got it? And the only thing I could think of was that it was more definite suddenly. I thought I had probably got it but I hoped I hadn't, and then 'you have', and so it's a shock isn't it, and I didn't realise. It was actually a physical shock when they told me you see.

Interviewer: So for how long had you been thinking that you might have before that?

Helen: About a year I think. Yes, there were things that I thought 'I know', I felt, my mother's feet twitched you see, which was the first thing that happened to her, and I thought 'ah I know', because that's what happened [to me as well]. (Helen, Interview 4, 13.12.2010)

For those who were unaware of HD in their family, denial and disbelief were common reactions to the news of their diagnosis. Depression and low mood

often followed during this time, challenging both the newly diagnosed person and their family.

When it was first, when I got the actual confirmation, I got it from my GP, and they (her children) were both there then. I mean we were so sure that it would be negative. We'd planned to go for lunch to celebrate and we just couldn't believe it. Just could not believe it. ...But we kept saying 'no, no it's negative'. You know, because it's not in the family, that's all we kept saying. But obviously it was. (Erica, Interview 1, 14.03.2008)

... for a long long time, couldn't accept it. Which is understandable in a way, you know it is not an easy thing to have to come to terms with and to accept. But that made it very hard for me because he wouldn't accept that he had got it. 'I am sure they have made a mistake' he kept saying 'there is nothing wrong with me, it's nothing like that'. (Margaret, Harry's wife, Interview 1, 17.10.2007)

These initial emotional responses to diagnosis were common in all participants' stories. Although early in the disease trajectory, each of the participants included in this chapter had signs of physical impairments, such as unsteady gait, changes in the pitch, tone and rhythm of their voice, and uncontrolled movements. Participants did not solely focus on the emotional aspects of coming to terms with the reality of their diagnosis. Each then went on to talk about some of the more functional impacts their diagnosis had had on their lives.

PRACTICAL CONSEQUENCES

At this early stage participants noted issues around slowed cognition, a reduction in fine motor control and some behavioural changes. Within a relatively short space of time these newly diagnosed participants must not only come to terms with their diagnosis but also with challenging life changes like giving up work (Tom and Elizabeth), or relinquishing their driving licence (Helen, Tom, Harry and Elizabeth). Depending on how early they were diagnosed, these events often happened within a few years and affected

adaption and coping mechanisms. They also had a wider impact on the family and their way of life. For example after a few months into the study period Tom was made redundant. The household therefore suffered a loss of income creating numerous financial and social impacts for the family. It also created a shift in the dynamic of the family when Tom was no longer working and his wife Nicola needed to continue to do so, as well as providing additional care at home. Tom's role in the household had shifted as he was no longer able to contribute in the ways that he previously had.

You wouldn't think that HD would cause so many little problems, do you know what I mean, just normal simple day-to-day tasks for a person that's not got it, it's quite, I just can't believe how much it does involve you that you can't do. ... It's like the DIY, like you struggle with that now don't you. So I've had to sort of take it on board obviously with Tom's saying 'do this, do that'. ... It's just that I still like to involve him because it must be so hard. (Nicola, Tom's wife, Interview 1, 21.10.2008)

As Tom was in his 40s and only in the early stages of HD the family had concerns about how he and his family would cope when he stopped work. However, despite missing some of the social aspects, Tom reported feeling relieved by no longer having to work. He recognised that over the past few years he had begun to find work increasingly stressful as his condition caused him to be less agile both mentally and physically. In order to keep busy and give purpose to his day Tom played golf a few days a week, used several interactive sporting computer games and arranged a rota with his wife and daughter to pick them up and take them to work. When Tom was encouraged to stop driving after a minor accident he adapted quickly and the family got a dog to keep Tom company in the day and active through walking. My observation notes reflect the concern expressed by the CNS for Tom and Nicola during this key transition of stopping work.

The CNS asks Tom if he has had any falls, he says he has not had any, says he is playing golf, no problems and thinks he will play more golf when finishes work. Tom says he has had enough of work, he has been there 25

years and doesn't feel valued. The CNS says we have time to plan as it could be September/October time when he finishes, but 'we need to get you into a routine'. The CNS suggests the rehabilitation support worker has a look for what else there is to do in the area 'if you get out of the routine you will get bored', need to replace it with a structure. Tom says that at the moment a rest seems a good idea ... The CNS persists and says it is time to think through how to replace work, she explains to Nicola about apathy. (notes on CNS and RSW home visit to Tom and Nicola, 15.07.2009)

Just over a year later I had arranged to see Tom and Nicola again for their second interview. Nicola said she was taking the dog to the vets to give me a chance to talk to Tom on his own. My notes from this interview reflect the changes in Tom since stopping work.

Much more chatty than has been previously, seemed more comfortable and confident. Talked about their holidays and finishing work, generally doing very well he doesn't seem to have any concerns and is pleased to have finished work now and is keeping busy and active. (notes on Interview 2 with Tom, 11.08.2010)

Elizabeth also reported being relieved to have stopped working. She recalled finding work very stressful over recent years as she struggled to keep up with a job she had been doing all her working life. She found trying to complete tasks at work placed strain on her slowed cognition and she struggled to multi-task, organise and plan her work load, making working an additional stressor.

And I've retired early because of Huntington's so I'm just beginning to enjoy life really ... I've been a (role) all my working life, and I've enjoyed that, but the last few years became a bit of a struggle really. ... But I feel so much better now I'm not at work. ... The last couple of years were awful ... I didn't feel I was doing a good job but I didn't know why. ... It was concentrating and doing work, I couldn't. Well I could do simple things but I couldn't plan and keep to it. (Elizabeth, Interview 1, 30.04.2009)

A couple of years later, towards the end of the study period (2011) Elizabeth also had to stop driving. Initially, she was extremely upset by this news but, like Tom, she was able to turn the situation around and find other ways to maintain her independence. Elizabeth had since discovered taking the bus to where she wanted to go. As a woman in her early 60s she enjoyed being out in the day with the other 'oldies' and regular bus drivers who were considerate and friendly helped her rebuild her confidence to be out on her own.

Interviewer: But you were quite upset at first were you?

Elizabeth: Yeah very, I was absolutely devastated, I cried all that day.

Interviewer: A little bit of your independence.

Elizabeth: Yeah. ... And confidence went. ... Yeah, but I got over it.

(Elizabeth, Interview 3, 21.07.2011)

Participants considered relinquishing the ability to drive as a dramatic dent in their independence. A number of participants noted stopping driving or having their licence revoked as a pivotal time (Tom, Elizabeth, Harry and Helen). Most were able to accept this change without too much disruption after the initial realisation. This seemed to be even more apparent for participants with family carers who could drive as they were able to take on this role and the family were able to continue with the majority of their activities. However for Helen, who lived alone, it was evident that being able to drive was a key practical aspect of her life essential for shopping, getting to church and social activities. She struggled with being reliant on friends, neighbours and taxi services for transportation. She very much missed being able to drive herself and this was raised in all her interviews.

It's been hard. ... I miss my driving. ... I miss the freedom of having my own car. I've been driving since I was 17 ... [The CNS] said it, losing your car is like a bereavement in a way, because you're so used to it, I love walking but I can't get very far nowadays, you see. (Helen, Interview 3, 18.02.2010)

During these early stages participants diagnosed with HD started to make small adaptations to their behaviour and environment to accommodate their HD symptoms. In particular several of the participants in the early phase talked

about problems with fine motor control and balance which forced them to adapt the way that they dressed, did the cleaning or other household tasks.

Nicola: You struggle with your buttons don't you on shirts which has been sort of an ongoing thing anyway. He's got to learn to sit down though to put things on. Like when he's getting dressed in the bedroom, he thinks he can still stand and put his underwear on, and he can't balance, and I say just sit on the bed. ...

Tom: Your brain tells you that you're still upright.

Nicola: I mean he has come to terms with it ... but it's like him helping himself more now, sitting to put things on and just little things like that.

(Tom and his wife Nicola, Interview 2, 11.08.2010)

FAMILY CARER RESPONSES

Family carers also needed to make adaptations to their behaviour to accommodate the needs of the person with HD. This did not happen easily and many talked of symptoms causing friction within the household. Harry's wife Margaret found both the physical and behavioural symptoms difficult to cope with. The couple had married later in life after their previous long-term partners had died and were looking forward to an active retirement together. Margaret felt frustrated that Harry's personality and physical changes had forced her to alter these expectations.

Plus this aggressiveness and nastiness. And he was really very nasty to me and said some horrible things over the time. ... but it's something that you have no control over. So although it was very difficult, at least because I had read a lot about the illness and realised that it was one of the symptoms, I could accept it. Whereas before [Harry was diagnosed], I was so worried and things were changing. And until I got the confirmation [that he had HD] I just thought well you know where do we go from here? I'd even thought along the lines of we'd be splitting up because I didn't want the new Harry. I wanted the old one back, that I'd married. ... So I don't get the opportunity to do things that we did together. (Margaret, Harry's wife, Interview 1, 17.10.2007)

Education for family carers was provided by the local support groups and the health professionals working in the area. After Tom's diagnosis his family struggled to understand and cope with his symptoms. Nicola became frustrated when Tom broke crockery and was not able to contribute to the household as he had previously done. However it was communication between the couple that was a particular issue. The couple sought support and information from the CNS to help them to better understand the condition. Nicola also attended an information and support day provide by the local HD team. She recalled how much it had helped her to understand her husband's condition, in particular his slowed thinking.

They got a gentleman in whose wife's got Huntington's ...and he was trying to demonstrate being in a Huntington's mind, and he was really good how he did it. It was absolutely fantastic the way that it came over. So it does make you stop and think because Abigail gets quite agitated with her dad when he doesn't answer straightaway. ... But if they said to them what colour's your top and where did you get it from they can't cope with two questions. So I say to Tom what do you want for your tea?, and then I go off in the kitchen do something else and wait until he's decided. So it's like working together in a lot of ways. ... Tom's mum came with me. So she understood it more. (Nicola, Tom's wife, Interview 2, 11.08.2010)

Like Nicola some participants specifically sought information and support in order to increase their knowledge and understanding of the condition and their genetic family history. However, this was as and when they wanted and was prompted by the particular issues they were facing. Generally information was sought from and supplied by the health and social care professionals involved in their care. In some instances leaflets written by the Huntington's Disease Association were used.

KNOWLEDGE MANAGEMENT

People's knowledge of HD prior to diagnosis was extremely limited. Some people inevitably turned to the internet for information but found this more frightening than helpful. Many had not heard of the disease and for those that had, any further knowledge was only basic. Even most of those who were aware of HD in their family had a limited knowledge of the disease. They attributed this to a general lack of knowledge among the health and social care professionals they had previously come into contact with. Both Rose and Helen noted that since their parents had lived with the condition many improvements in knowledge and treatment had taken place. Prior to genetic testing diagnosis was not always accurate and therefore treatment, management and information could not be optimised. Some participants had stories of previous generations being 'put away' or 'sent away' to mental health institutions and sometimes prison when there was not the knowledge to manage the disease in the community. A silence was often created around HD. So the diagnosis was known but further information seeking was not encouraged or may be blocked, creating a tacit agreement not to further discuss the illness or its implications. Not talking about HD was what some respondents referred to as being 'brushed under the carpet'.

When mum had got it, my dad went [to the consultations], I didn't. I was the younger girl, so you didn't. It wasn't, everybody knowing. ... It was brushed under the carpet. (Rose, Interview 1, 22.02.2008)

Harry and Elizabeth also reported poor transference of knowledge from health professionals during their diagnosis. When diagnosis was unclear, people undertook a barrage of tests, the genetic blood test for HD was sometimes included within this assortment. Some families had been warned that HD was a potential diagnosis, although as Harry and Margaret explained it was not always clear the genetic test was being undertaken to confirm this;

He said 'I have a good idea of what I think it is' So he said well there is just one final test we have got to do to confirm it, that's theblood test. But even then he didn't say it was a gene test. But it must have been ... And

*he said I will see you in a month, he said and we will tell you for sure.
(Margaret, Harry's wife, Interview 1, 17.10.2007)*

Elizabeth and her husband did not recall being given any indication HD was being tested for and no warning of the potential outcome and the implications of the positive result. The continued distress caused by the way they were informed of Elizabeth's diagnosis was evident throughout the couple's first interview.

James: But no, we had no thoughts at all about it, we were not informed, we were not told. We might have been told that this test is being done for blah, blah, blah, but that was it.

Elizabeth: You don't take that seriously do you?

James: it was just a tick sheet really, that this will test you for blah, blah, blah, blah, and that was Parkinson's, Huntington's, but it was never mentioned that this was a serious possibility. (Elizabeth and James, Interview 1, 30.04.2009)

INFORMATION SEEKING

Once diagnosed all the participants in some way explored their family history in order to identify the origins in recent generations and movement of the disease. Complex family dynamics, early deaths from other diseases, later onset in relatives and significant age gaps between family members all contributed to probable but uncertain mapping of the disease in some families. For Tom the premature death of his biological father from suicide meant that he had been raised without contact with his biological father's side of the family. However further enquiries to his mother revealed the suspected traceable line in the paternal side of their family.

Abigail: But granddad is not your real dad is he, so that's why we're not sure.

Tom: This is what we suspect.

Nicola: They're 99% sure it's come from his biological dad. But he actually committed suicide in his thirties, so we can't sort of trace ... But from what

his mum's told us because she got married at sixteen and had Tom, and she actually lived with Tom's dad and (his) dad, but there was no [grandmother] around, and then [Tom's mum] said they never really spoke about her but the times that they did she was in (specialist) hospital. So she seems to think that she had HD because apparently they used to put them in mental homes years ago because they thought you were mental. So she's sort of like reading between the lines. (Tom, wife Nicola and daughter Abigail, Interview 1, 21.10.2008)

Misdiagnosis also contributed to a lack of understanding in some families. Participants reported several cases of symptoms being confused with those of Parkinson's disease. Due to the late onset of the disease in Rose's family it was her uncle that was misdiagnosed with Parkinson's for a number of years until her mother was diagnosed with HD at the age of 58. Similarly, due to late onset in the family, the symptoms displayed by Harry's older sister, who lived to an age of 78, were attributed to dementia.

CONTROLLING INFORMATION

Some respondents talked about their own silence and the reasons why they had or had not chosen to tell people in their social and work circles. For some this allowed them some time to become accustomed to the diagnosis; whereas for others it was a conscious decision to maintain their privacy.

I think initially you tell too many people, because you just blurt it out. I have blurted it out. But again on [the RCA's] advice it's 'well if they don't need to know don't tell them.' So we're not doing that. They can think what they like, we're not bothered about that. ... If they ask I'd tell them, but nobody around, no neighbour knows, it's just close friends that know and family, that's all so. (James, Elizabeth's husband, Interview 1, 30.04.2009)

So, but one or two people had noticed I was wobbly. ... It's one of those things I feel quite strongly about. I don't want everybody knowing about it. Because they all start looking for symptoms don't they? (Helen, Interview 1, 11.12.2007)

The symptoms of HD can often be misunderstood for other things and a particular concern for some was that they might be perceived as drunk when their gait was unsteady or they are unable to control movements. However one family participant wanted to control information by being the ones to inform people directly. They felt that in order to overcome this type of stigma they needed to be open with people about the condition. In their first interview Tom's wife Nicola, and 21- year-old daughter, Abigail, were both present. They were a close family and often went out together for meals and to their local pub.

Nicola: I mean Saturday night we went out and he went to get off the stool in the pub, and he stumbled, and there was this couple sat near us and he sort of looked at him as if to say 'oh a few too many (drinks)', ...

Abigail: We've had a few people say that, haven't we, when he looks like he's had too much to drink.

Nicola: And he says to them 'I'm not drunk mate, I've got Huntington's disease', and he went 'no problem'. ...Yeah and I think that's better to make people more aware. (Tom's wife Nicola and daughter Abigail, Interview 1, 21.20.2008)

Elizabeth and Helen also talked of using a walking stick to act as a visual sign to strangers that their unsteadiness was due to a medical condition rather than bear the stigma of being viewed as having had too much to drink. I would suggest that it is also possible that these participants cited stigma as a social excuse for having a walking stick rather than acknowledging that they might need support and assistance when walking.

INFORMATION BLOCKING

Despite most people using a range of systems for information seeking they also utilised complex systems for blocking information. Participants and families reported a number of ways in which they limited their own and other peoples' knowledge. For example a number of participants minimised or denied symptoms in order to put off finding out their cause. Margaret reported being concerned about Harry's symptoms for a while prior to his diagnosis. However Harry accepted many of his symptoms such as memory impairment, and lack of

interest in activities as a part of advancing older age. However being seen for a routine flu vaccination at their GP surgery prompted the nurse to suggest a fuller examination, as Harry's wife, Margaret explains;

And we actually went to the doctors for a flu vaccination didn't we. And the nurse said to us ... 'don't you think it would be an idea to sort of have a check with your doctor?'. So we made the appointment ... Because I thought good, this is really what I was hoping, somebody else might pick up on it because I didn't quite know what to do. So we made the appointment. ... And as soon as you walked in he (the GP) said to you 'how long you been suffering with this jerkiness?' And he sort of picked up on the fact and started asking you a few questions. (Margaret, Harry's wife, Interview 1, 17.10.2007)

Some participants suggested that the shame, stigma and guilt attached to the diagnosis had, in previous generations, prevented information and knowledge being gained and passed to family members. Helen's mother was never told she had the disease to prevent her from feeling guilty about potentially passing it on to her daughter. Helen's mother was identified as having HD in the 1960s and died in 1980, however initially doctors were not clear if she had HD and by the time it was officially diagnosed discussions were predominately held with Helen rather than her mother who was often heavily medicated. During this time professional knowledge of the condition was limited. Both the GP and neurologist had never seen a case of HD at this time. Hence as her mother's condition worsened she was never told she had HD and health professionals were instructed to maintain this secret.

Helen: Again most people (health professionals) who would come round like that, I mean I told, I wrote [the GP] a little note because I didn't want my mother to know what she'd got, because of it being hereditary.

Interviewer: Oh did she never know?

Helen: No. She knew she'd got something but we called it something else. Because she, I knew she would have been, well she was brave with her own

illness but if she thought she'd passed it on to me, she'd have been really upset. (Helen, Interview 1, 11.12.2007)

Breaking this silence seemed to happen only over a number of years.

Participants talked of the strain a diagnosis of HD could place on families. Some participants talked about family conflicts that arose and have been, or, are being resolved. However, for some families living with HD over the generations had caused rifts and separations that meant people had family members they have not had contact with for a long time. Rose explained how her sister removed herself from the family when their mother's disease became apparent, and had not made contact for over 20 years;

And she took off and didn't want to have anything whatsoever to do with this at all. She wouldn't visit my mum or my dad or anybody. People were, it affects everybody different doesn't it? So, I've not seen her since. (Rose, Interview 1, 14.03.2008)

This disengagement from the family was the most extreme form of information blocking. Others reported instances of family members refusing information or to engage with conversations about the condition.

I mean at first when I told (my brother), he's a bit like he doesn't want to know, you know what I mean - he'd say no I can't, I'm not being tested, I'll just wait and see. It's like he's obviously had a word with his wife and it's like we've said he has to know because he's got kids and grandkids. But luckily he's okay, it's just me. (Erica, Interview 1, 14.03.2008)

Not wanting to know could be considered a form of coping. Blocking this kind of information was a common way for family members to deal with the genetic risk they now faced.

'NOT WANTING TO KNOW'

A confirmed diagnosis of HD has implications for all those related to the person diagnosed and the impact on other family members was discussed by some of the participants. Nine of the total fifteen participants had adult children and five

had grandchildren so the implications of their diagnoses were far reaching. For a number of the participants' family members the decision to be tested themselves was complex and difficult, particularly if they already had their own families (genograms in the individual case profiles also depict family members who are positive for HD, HD free, or remain at risk). One element that seemed to prompt children of participants to talk about being tested was if they were planning their own family and wanted to make an informed decision about whether to have their own children. Harry had a large family with three children of his own, none of whom had been tested during the study period, partially because they currently had no plans for family of their own.

They say at the moment they don't anyway. I think they would rather not know as things are at the present time. ... I suppose job prospects, insurance, there are all sorts of things that come to mind, that you can understand them putting off and not wanting to know. ... So I think they both (his sons) said if they wanted to settle down with somebody then they would feel they probably needed to know. ... his daughter is married. But they have not wanted children anyway. (Margaret, Harry's wife, Interview 1, 17.10.2007)

Some participants had relatives who had since been tested and were free from the HD gene. However participants talked about a number of relatives who had made the decision not to be tested at this time, preferring not to know their status. Erica spoke of how difficult the decision was for her two children, both in their late 20s;

They keep changing [their minds]. Well [my son] is one of them, I don't know, he won't talk about it. At the minute he's said no. I mean at first they both said yes they did. Then they both changed their mind. But [my daughter] said - I mean there's been a lot going on in her life, I mean with me, I mean we lost my mother-in-law last year as well, ... And she said at the minute it's not right, but maybe next year. (Erica, Interview 1, 14.03.2010)

At the age of 21 Tom's daughter, Abigail, explained her reasons for not wanting to be tested at this stage in her life. Her age and the fact that she did not intend to have children of her own at this time was a factor in her decision not to be tested.

Abigail: There's the test that I could have to know if it's done, but I'd have to go through all the counselling and I don't think I want that really. I just want to get on with my life.

Tom: She's not thinking about having a family or anything yet, hasn't got a boyfriend or anything, so there's no point is there really, it's up to her.

Abigail: I don't really want to know about it at the moment. ...

Tom: Probably if you're thinking of having a family or settling down then it's different. I mean she's not bothered are you?

Abigail: Not really, no I just want to go out and party. (Tom and his daughter Abigail, Interview 1, 21.20.2008)

For those children who already had their own children, passing on the gene could not be reversed therefore they argued that getting tested posed little advantage for them. Helen had no children of her own but Rose recognised that she had made the decision not to be tested when her mother's HD was confirmed as by that time she already had her own children.

But I mean they're all grown up now. My son in Ireland, he's got two children, five and eight, and he's not having the test either. I can see where he's coming from, because I did very much the same sort of thing. (Rose, Interview 1, 22.03.2008)

Although Rose construed her son's actions as the same as her own there is some difference as he knew of his grandmother's diagnosis and therefore his own level of risk even without his mother being diagnosed.

BALANCING SERVICE INPUT

The health and social care professionals took on a number of roles during these early stages, firstly to help the person and their family come to terms with their diagnosis, and then to help them to maintain their current health status for as long as possible. The health care professional involved in this study recognised there to be generally three phases or stages to the course of the disease. All noted that the different stages required different types of input but that the early stage could be the most challenging for health professionals needing to make judgements about how and when to provide support, advice, information and additional care services.

We have what we call levels of activity with each of those. Level 2 is the main part, which is about people that are becoming more symptomatic, have got perhaps behavioural physical problems, carer support, placements for respite, that's the biggest part of my work. Level 1 are newly diagnosed, which is more about advice and support to them, and Level 3 is usually palliative care, really people at end stage or at the last stage of the disease. A lot of them are in residential home care, but I do a lot of teaching there so my role changes within that environment. (CNS, Interview 1, 24.06.2009)

The RCA noted that the early phase of the condition in particular required more intricate and creative help in order to engage with patients and families.

Often I think that the earlier on in the condition where you're still adjusting is actually the hardest, and I think it's often that they have more problems ... So in fact it's at that stage where they need perhaps the most intricate help really and the most creative help because how do you help someone who is perhaps not really acknowledging that they are really struggling, and how do you weave it in to their thought processes without them putting down the shutters and actually [not] letting you near them. (RCA, Interview 1, 20.10.2008)

The health and social care professionals highlighted that the early phase could span a number of years and sometimes decades after their first encounter with the services. Both the RCA and the CNS had been in their respective roles for a number of years and were able to provide a level of continuity that could be considered rare in health care. Both noted the importance of getting to know the person with HD and their family in order to make judgements about how much and what type of care input might be the most appropriate.

So it's continuity and building the confidence of people, knowing what services are out there and accessing it for them, so you're like an advocate too, and that raises lots of issues from gaining access to Social Services care right the way through to employment. ...So it's about supporting ... and signposting them to the right people to help them ... it's all facets of their life really that you get involved in, because that's what my role is. (CNS, Interview 1, 24.06.2009)

Other health professionals involved in the study also noted the importance of familiarity and continuity of care. The speech and language therapist explained how her skills may not be needed during the early phase of the condition but that she met all patients attending clinic to build familiarity with herself and the service early on. She felt that at the point her input was then required this was accepted more easily by patients. Until this rapport and knowledge was established health professionals struggled to make decisions about how to balance their contribution and their involvement. The newly diagnosed person was most likely not well known to them hence judging how much input and information to give was a challenge for the key health professionals.

I also think that actually if you haven't heard, it's about how do you look at the situation if you haven't heard from someone is that a bad thing? That might be a good thing, getting on with their lives. So it's a difficult one, it's all about timing and knowing, knowing the family, but I'm hoping that certainly I know them well enough that they would contact me and they've felt that they could. But again I think you can't always rely on that because

I probably know myself that sometimes I might, I daren't ring somebody in case, there's always that oh I don't want to be a nuisance, and I hope that people aren't sitting at home thinking they are a nuisance [and are afraid to ring me]. (RCA, Interview 1, 20.10.2008)

All health and social care professionals utilised home visits and clinic appointments as ways of assessing their patients. This information all contributed to the knowledge built up about patients and their family carers.

...it can give away such a lot. You can look at a person's well-being by the state of the house but you can monitor them over a period of time and when you start to see things are not quite right... (CNS, Interview 1, 24.06.2009)

My fieldnotes on an observation of one of Harry and Margaret's clinic visits show how these assessments continued in the guise of general conversation.

We are sitting in the waiting area having just seen to the consultant, the CNS comes over to say hello as she was not in the consultation as usual. She started with some general conversation and then focused on asking Harry some questions about football and a recent local game to test memory, he is able to recall most of the answers without help and the rest with a little prompting. She encourages him to keep his mind active by trying to recall these kinds of questions. I am now recognising the ways in which the staff ask questions to test memory, speech and for confusion. (Notes on observation of clinic visit with Harry and Margaret, 28.01.2009)

Helen was one participant who recognised that her clinic appointments and home visits served a number of purposes and that a level of surveillance was part of her routine care.

When [the CNS] comes she generally has a chat to me to see what I've been up to, make sure I'm active. They like to keep you active - still doing your crossword, yes, yes, in the paper, and that sort of thing. Note the sort of things I've done at church, because that's how you're social and you have to

be talking quite a lot there. So she'll always do that first and then she can tell if I'm still the same or if I've got any worse. ... It sounds just like chatter but it isn't really because it's important that she can tell whether I've got any better or worse. (Helen, Interview1, 11.12.2007)

The level of input also needed to be balanced with the type of input. Health and social care professionals were aware that the way in which they responded and behaved in clinic appointments and home visits would be picked up on by patients and family members. In the extract below the RCA is talking about how Elizabeth's husband James had changed his attitude after the initial devastation of Elizabeth's diagnosis. The RCA grappled with concerns that positive responses from herself and the consultant in clinic may have prevented James from seeing key changes in his wife's condition, yet this change in his attitude had enabled them to take positive steps to continue their life as 'normal' and go on holiday.

It was very interesting, James, he literally was watching her, watching every every little twitch, movement and saw Huntington's as completely catastrophic. And had a very catastrophic vision really for the future, that everything, that his life and her life, that's it really. And then when we last saw her in clinic it was almost as if 'she's fine, she is fabulous, she is doing this, this'. Where as we then saw huge changes, so he has gone completely the other way now, he is almost not even seeing any. ... But the change is quite remarkable where he is almost not getting it now. So I don't know where, what has changed, what's gone wrong, who is being over positive, have we? But you know how sometimes, certainly in our clinic, you must have seen it where we perhaps over do the positive side, because how can you sit there, you can't mirror the bleakness, and maybe sometimes we are over [positive], I don't know, it's a real food for thought actually. Generally it works, in a way it sort of worked, because they both saw their lives coming to an end and now they are going on holiday. (RCA, Interview 2, 07.07.2011)

LEVEL OF INPUT

During this early phase, maintenance or improvement of current health status is the focus of care and unless otherwise sought participants described being left to continue their lives as they wished. Professional input was provided at yearly out-patient clinics. This yearly check-up or 'MOT'⁶ was a way for both service users and providers to keep in touch. Providers used these appointments to reiterate their availability, and to identify any potential or current issues which they could help resolve. At the out-patient clinic in Birchtree I observed each patient routinely being seen by the consultant, dietician and the speech and language therapist during one appointment. The CNS would also be in attendance, co-ordinating the clinic and sitting in with the consultations if necessary. These fieldnotes reflect the types of consultations that occurred with the dietician over the study period as she monitored their weight and eating habits.

The dietician weighs Helen and says she has put a little bit on which is fine. ... The dietician asks her to run through what kind of food she has in a day, she focuses on the amount of milk she is using, only about a litre a week. The dietician explains she is trying to find food with calcium in (demeanour is generally friendly, encouraging and remembers things from previous consultations which make her sound interested in the patient as an individual). ...[the consultation continues]. The dietician says she is happy as Helen is eating well, so she is happy to just see at the next clinic. ...the dietician says 'you have got my number, even if it is not the HD side there are others in the office who can help so no problem to ring'. (notes on observation of a clinic visit with Helen and the dietician, 29.09.2010)

In Birchtree the CNS explained that she took care to organise clinics to cluster those at different disease stages. She noted that experience had taught her that

⁶ In England cars over 3 years old must undergo an annual MOT (Ministry of Transport) test to check that the car is road worthy. Patients used this as a metaphor to refer to their annual consultation appointment to check on their general health and any potential disease progression.

those with early disease could be frightened by seeing those with more advanced disease at clinic and in some instances may be put off attending appointments. Patients also recalled that they did find visiting the clinic frightening and stressful at first.

That was my first visit [to the clinic] last year. But I had six months off. Because I thought well it gave you the shock of going. Why not, so I asked them to leave me alone for six months and they did. (Rose, Interview 1, 22.02.2008)

Other participants needed more intensive input to help them through the period of coming to terms with their diagnosis and finding their feet in a new world of health and social care services. Below are extracts from fieldnotes in Harry's case. They reflect my observations of some of the CNS home visits with Harry and his wife Margaret. Much of the focus was on getting additional support for Margaret so she could continue to help Harry with his daily needs. During the summer of 2008 the CNS was visiting the couple on a monthly basis and I attended three of these home visits. I wanted to attend this visit to see if Margaret had followed up on a previous suggestion to have a sitter so that she could leave Harry for longer periods.

The CNS asks Margaret 'have you thought any more about what you might need because we are not going to let this drop' (worded in an encouraging but forceful tone). 'What else can we do to support you?' ... The CNS suggests a local sitting service again, and says 'think about it'. ... Margaret tells her she tried a local number but wasn't able to get through and thinks it is a lot of effort. The CNS asks if she would like her to pursue it for them? She redirects this to Harry, he says ok, the CNS says 'if we don't try we won't know' (uses 'we' not 'you' – less accusatory and more team like). ... Wife talks about getting appointments for her own illnesses the CNS just listens and offers some basic advice (Talks for quite a long time, the CNS let her vent and feel heard). ... The CNS makes appointment for next visit in one month's time. (notes on observation of CNS home visit to Harry and Margaret, 23.06.2009).

At her next visits one month later the CNS followed up again on this issue of support for the couple. Margaret reported that the CNS had met with the whole family to discuss Harry and Margaret's needs. Since that visit Harry's adult children became more involved in his care and in supporting Margaret.

During these early stages a key part of the professionals' role was to provide information. The Huntington's Disease Association was a primary source of information and the RCA felt much of her work involved giving information and correcting misinformation. In this extract she is referring to Elizabeth and James' case.

I still think the key role, the aspect of my role is information giving and actually putting right misinformation. I mean even the family that we had in today. When they were told they were given news about Huntington's in a very cold way, it was through a letter and they weren't really given information, and then the husband went online and then found out all the worst possible scenarios that can happen and thought well that's going to happen like next week. So it's putting things right, or trying to. (RCA, Interview 1, 20.10.2009)

As identified earlier in this chapter many participants struggled to come to terms with their diagnosis and relied on the CNS and RCA for support, information and advice during this time. In two interviews with the CNS she recalled her input with Tom and his family. After Tom's diagnosis she was arranging regular home visits every few weeks and follow-up telephone calls to help them come to terms with the diagnosis and what it might mean for the whole family. Input was for Tom himself, his wife, their daughter and Tom's mother and step-father. The CNS saw them together as a family but also met with Tom on his own and his wife Nicola on her own to establish and support their individual needs. A couple of years later the CNS noted how her input had decreased as the family accepted the diagnosis and were coping well with the changes it has brought them.

So with Tom it was shock and anger we had to deal with. With his wife it was complete distress. She tried to be very strong about it but was actually

frightened, what does this mean? And also for his mum and extended family, it's supporting them. So a lot of my time was spent with measuring Tom's acceptance of the diagnosis, also monitoring his level of depression, because I felt quite soon on he was a very depressed man, so treating that. Supporting the wife, I've done sessions with her on her own and also sessions with Tom and with them together. (CNS, Interview 1, 24.06.2009) ...about 12 months ago I was visiting quite regularly just to keep tabs on them because mentally he wasn't doing too good and it was causing friction in the family. But that seems to have subsided so [now I] leave them alone, they are ticking along quite nicely. (CNS, Interview 3, 27.06.2011)

The combination of home visits and clinic appointments allowed health professionals to maintain contact with patients and carers. However, outside of these encounters they were also reliant on patients or family carers making contact or asking for help when it was needed. All reported contacting health and social care professionals for support, advice and information. Helen noted in her interviews that she would occasionally ring the CNS between her six weekly visits if she needed information or just someone to talk to. Those living alone, like Helen and Erica were of particular concern for the health professionals. Balancing the right level of input with this group of patients was more challenging. In this instance the CNS is talking about working with Helen.

As you know she comes to clinic, that is still ongoing and that will continue, that's probably going to increase over the next couple of years to 6 monthly at least. I'm seeing her about every 4-6 weeks. ... One because she is what I would say would be at risk because she is on her own and her age, not so much the Huntington's but because she is on her own. Those type of people I find can cause more difficulties if I am not keeping an eye on them and getting things in place sooner. If there is a carer involved they are more likely to alert me, and they are more likely to comply with what I suggest. When they are on their own you have to really keep working with them. You can't suddenly appear and 'right I think you should be having this now', you need to be able to chat with them about that. ...So it is that fine line isn't it between you don't want to make her over dependent but there

*again you need to be able to keep an eye on her. So it is that fine line really.
(CNS, Interview 2, 14.12.2010)*

Due to the differences in the CNS and RCA's roles such close monitoring was not available for Erica.

A key element for both carers and health and social care professionals was to help the patients to remain as socially, mentally and physically engaged as possible for as long as possible. Lack of motivation is a common manifestation of the condition so it was important for family carers and health care professionals to have strategies in place to help maintain their functioning. This involved a wide range of professionals including physiotherapists, occupational therapists, and in Birchtree, the rehabilitation support worker. In this interview the rehabilitation support worker is talking about his input with Rose and her daughter Charlotte.

I encouraged her out and into the gym, ... we brought all these things in and it did lift things for a little while, but then they seemed to slide back, ... like the cooking that was another thing. I think Rose had burnt herself at some point, so Charlotte had stepped in and was taking over more in the kitchen, but ...it's good for you to keep doing stuff and we can look at what you're doing and make sure if it's just about not using heavy pans then you can work together and Charlotte can do that bit and you can do other stuff, but to eliminate that out of her life it puts a lot of pressure on Charlotte and it deskills Rose (Rehabilitation Support Worker, Interview 1, 29.03.2011).

The nature of HD means that it is harder to create new memories so the health and social care professionals recognised that people would often return to learnt habits and ingrained memories. The rehabilitation support worker noted that he had provided consistent input by returning to support Rose approximately every three months to maintain her levels of activity. The rehabilitation support worker also supported Harry and his wife by spending time taking Harry out for walks so as to increase his confidence and physical fitness. It also meant that Margaret did not have to feel like she was nagging

Harry to keep active and allowed him to have some male company. Helen had input from a physiotherapist to help maintain her core stability for balance and walking. She felt that having this physiotherapy had reduced the number of falls she had had.

THE KEY WORKER

Both the CNS and RCA were considered important sources of information and support during these early stages of the condition. Their disease specific knowledge and advice was highly praised and considered to be an essential aspect of care by participants. They were also able to direct people to other services and could act as advocate for families when needed.

I think it is how she comes over with things, and her knowledge of it. I just feel that you ask her a question and she has got an answer for you. ... She makes you feel more at ease. It's not like you're at a desk one to one, she comes to the home, she really makes the effort to come out and see Tom and everything, which she does with everybody else I would imagine. But yeah, she's just there for you all the time. (Nicola, Tom's wife, Interview 3, 26.06.2011)

This accessibility was also an important aspect of the role, particularly for the CNS. She was considered to be central to care and the 'first port of call' for help and advice. However there also seemed to be something fundamental to her character that participants felt contributed to the role.

We can talk things through ... If I have one of my bad days I'll ring her up and perhaps just chat to her on the phone or if she's got time she'll do an extra visit but I know she's got so many people to see hasn't she. She's so busy I don't like to ask if I can possibly avoid it. ... she makes herself so available. ... She's got such a nice manner and she's so knowledgeable, and the two things together are great. (Helen, Interview 2, 22.09.2008)

Despite their accessibility a number of participants commented on how busy the CNS and RCA were, the large geographical areas they covered and the limits to

their services. All participants struggled with their entitlement to access services and tended to think that there were other people who needed them more. This meant that they tended to contact them in emergencies only. This was particularly apparent once they became familiar with the system and recognised the pressured nature of the CNS and RCA roles.

...it's just one of those things, she is so busy isn't she, that's the problem, when you ring her, she is busy. ...If I don't hear I don't worry about it. Mind you I ring her as well. (Erica, Interview 3, 03.11.2010)

Despite being considered available when needed the CNS and RCA were identified as a limited resource. The RCA role in particular was supplied by the charity sector and therefore limited in its scope. The role was one of liaison, advocacy and advice and therefore the RCA could not directly make referrals and did not have official standing with NHS staff. Recognising the CNS and RCA as limited resources created a tension for patients and families around when it was appropriate to contact them and their entitlement to services. However these people's specific knowledge of the disease and the individual often made them the sole key contact for families. This created issues when that person was not available such as during annual leave periods. However the small population of people with HD means that service providers may need to balance the funding of a specialist with the population of people served with HD.

I think, well if you ask your GP ... he doesn't know anything about it, does he? And that's the thing we always feel, nobody understands. If we've got a problem and we've got the [Birchtree HD] team, we can ask them you see. ... they have so much knowledge. I think that's the essential thing to get across from my point of view. And they understand you ever so well, and that's nice. (Helen, Interview 4, 13.12.2010)

So we were quite worried then because I found that if [the CNS] isn't around and isn't available then you are very very vulnerable and very very alone. You know, trying to get help or information from anybody else is pretty hopeless. (Margaret, Harry's wife, Interview 1, 17.10.2007)

The Birchtree team was aware that there was an issue of access when the CNS was not available. This was being addressed by the introduction of the rehabilitation support worker who was to be considered an additional key contact for the team and was able to take on some of the broader roles of the CNS. However, a dedicated NHS HD team may not be feasible in all areas even on a secondment or part-time basis making the role of the RCA essential to the system of care provision for HD.

BALANCING NEEDS

Balancing the needs of patients and family carers was raised by the participants although not by the health and social care professionals interviewed. Not all participants were completely happy with the service provision. One family carer particularly noted that she felt there to be a lack of services for carers. Margaret felt the CNS did not support her individually and that the focus was on her husband.

One thing I always felt was lacking in the HD service is that it's all geared up to the patient. ... It's not easy, it can be embarrassing and you don't want to look as if you're complaining or even showing your ignorance, because you don't understand things and you want a bit of help from a professional in how to cope with different situations. (Margaret, Harry's wife, Interview 3, 19.08.2010)

These comments did slightly contradict my perception of the service the CNS had provided to Margaret and Harry. In my observations of their home visits the CNS had taken considerable time to work with Harry and Margaret together. However, I also found that it was difficult in several instances to speak to family carers on their own. This was particularly an issue when the person with HD could not be left alone and did not go out alone, such as for day care. I was not able to interview Margaret by herself and it is understandable that she felt she did not get a chance to speak to the CNS without Harry present. However part of the reason for the monthly visits by the CNS during the summer of 2009 was to encourage Margaret and Harry to get services and support in place so that Margaret could leave Harry for a period of time. Harry was resistant to using

day care services which would get him out of the home and allow the CNS to visit Margaret alone if she wished.

This view is in contrast to Nicola's accounts who felt that the CNS had specifically provided a large amount of support to her and her wider family. As Tom was still working at the time of his diagnosis it may have been easier for the CNS to visit and provide support to Nicola without Tom present. Nicola spoke of a number of instances when the CNS had met with her on her own to discuss any issues she was having and had offered to speak directly with their daughter and Tom's mother and stepfather if they wished.

...[the CNS] comes out, if she doesn't come out to see both of us she comes out to see me, and we have a chat and - very very supportive. And I think you do need that. (Nicola, Tom's wife, Interview 1, 21.01.2008)

Throughout this period and during each of the phases an important aspect of care management provided by health professionals was to be thinking ahead, anticipating future care needs and potential areas of concern. This was particularly important at these early stages when people had little understanding of the disease and may not be able to think ahead for themselves. In this telephone interview I asked the CNS what sorts of things she was thinking about in terms of Tom's future needs.

...we are going to look at how long-term they are going to manage in that house. From the point of view of him accessing upstairs and the bathroom. But, like I said the OTs are involved, ... we have got to keep him as active as possible. That would be the only thing, is long-term he is going to need further adaptations or a level access home. He is very keen he wants to stay at home, understandably, and I think [Nicola] is quite happy to take all that on board as well and we will just put packages of care in because she has got to continue to work for as long as she can. So it just about, developing and organising and co-ordinating care packages as his needs increase. (CNS, Interview 3, 27.06.2011)

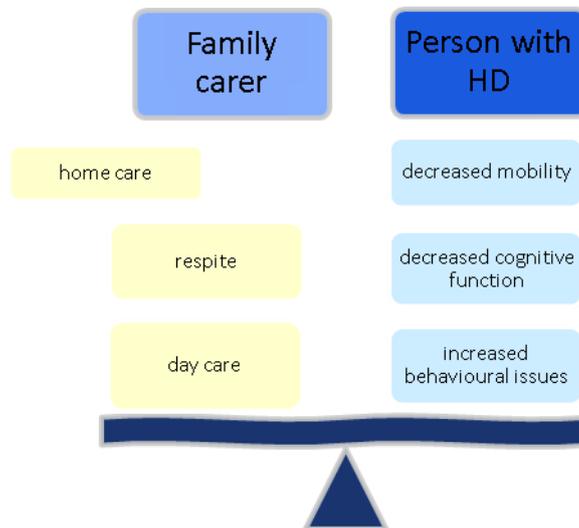
CONCLUSION

This chapter has presented findings associated with the early phase of HD, drawing on those cases which provide insight into these issues. Focus has been on how participants were diagnosed, and their response to this diagnosis. During the study period all were able to maintain their personal care and the majority of their activities of daily living, but did experience some key changes in terms of their cognitive functioning. These changes prompted those newly diagnosed with HD and their families to create of ways making emotional and functional adaptations. I have also highlighted the ways in which participants have balanced various forms of information seeking and blocking behaviours in order to gain, control and share knowledge about the condition. This knowledge management is examined as a coping mechanism in the discussion chapter.

The fourth section of this chapter drew on participants' experiences of service provision and the ways in which health and social care professionals balanced their input with the needs of patients and families at this early stage. They did this by helping people come to terms with their diagnosis by providing information, support and advice. As families settled with their new knowledge, the focus changed to maintaining, or improving current health for the patient and arming families with information to adapt to the changes in their loved one. Health and social care professionals tried to strike this balance so that they were supportive but not interfering. A key element of these early interactions was to build rapport with, and knowledge of, the patient and family.

Professional input was provided at yearly out-patient clinics and home visits tailored to the needs of the individual. Providers used these appointments to reiterate their availability as a resource to draw on, and to identify any potential or current issues which they could help resolve. How professionals balance the level of care provision is considered in the discussion. The role of the key worker is also taken forward in the discussion as a recurring element of service provision.

CHAPTER SEVEN: ACCOMMODATION AND MAINTENANCE



Mid phase - balancing accommodation and maintenance

INTRODUCTION

In the previous chapter I addressed the issues faced by people with HD and their families at the early stages of the condition. During the early phase people generally managed their lives on their own and remained in their own home. They required support in the form of information, advice, and signposting from health and social care professionals. Yet with support the person with HD remained able to carry out their own personal care tasks and most activities of daily living. The majority of this support was provided by family carers. The participants in this study remained in this early phase for a number of years and often decades (depending on when, in their illness trajectory they had been clinically diagnosed).

As the person with HD experienced decreased mobility, cognitive function and more complex behavioural issues there were increased demands on family carers and additional services were required to help maintain the person with HD at home. Again these phases must be viewed as ideal types rather than as having fixed boundaries. People did not fit squarely into a set phase, particularly

as their condition changed over time. For example, Sarah’s story started with her living at home, supported by a home care package, respite, day care and the 24 hour support of her husband. However after only a few months in the study her husband suffered a minor stroke and was no longer able to provide care for Sarah, prompting her to move to a residential care home, hence her story is more prominent in the next chapter. Amy, Martin and Vicky all had numerous complex needs and had care packages in place throughout the study period. In each case these packages of care provision were adapted over time. Sophie had no home care input but her personal care needs and activities of daily living were supported by her sister Melanie. As the study period progressed Sophie was beginning to need additional help and her sister and the Birchtree team were discussing how best to expand her care package to include some home care services. Table 11 shows the cases that inform this chapter.

Table 11 - Cases to illustrate trying to maintain balance

Name	Age at diagnosis	Time since diagnosis at start of study	Living situation	Services in place
Amy	27	10	Lives at home, is fully dependent on her husband, Ben and young daughter, Isabelle	Home care (morning and evening) Day care (two mornings/week)
Martin	55	5	Lives at home, is fully dependent on his wife, Julia	Home care (Day and night) Sitters (7 hours/week) Day care (3 days/week) Respite
Sarah	55	6	Living at home, fully dependent on her husband, Max and carers (moved to residential care after a short time in the study)	Home care (morning and evening) Day care (3 days/week) Respite
Sophie	40	7	Lives at home, is supported by her sister, Melanie	Day care (2½ days/weeks) Respite
Vicky	19	6	Live in an independent living facility, is supported by her mother, Sheila and brother, Jason	Home care (morning, lunch and evening) Personal assistant

This chapter is divided into five sections. First I look at how HD becomes a ‘space invader’, as areas of the home are increasingly dedicated to living space for the person and/or their necessary equipment. The chapter then goes on to

address the types of care services used by participants including day care, home care, and respite services. Not all participants welcomed this perceived invasion and the last part of this section on service input presents data from those resisting help from services. The third section focuses on the need for services to be tailored to the individual. Elements such as age, ability and home circumstances must be considered by care providers but also the specific issues posed by those with HD and the balance needed between autonomy and risk. The fourth part of this chapter addresses the bureaucratic issues encountered by family carers and health professionals. The final section then goes on to illustrate the challenges of caring, particularly for non-spousal carers and how some sought to achieve a life balance by aiming to maintain a degree of independence from their caring role.

SPACE INVADERS

Many families undertook adjustments to their homes in order to accommodate the facilities needed by the person with HD. Unlike the minor changes made to support mobility and independence in the early phase, these adaptations were more substantial. Both Amy and Ben, and Sophie and Melanie, moved to bungalows to allow greater mobility for their family member with HD. Amy and Ben were provided housing by their local council so when their previous house was no longer suitable they were moved to a specially built bungalow with wheelchair and hoist access. Despite not wanting to move it became a necessity after Amy had numerous falls down the stairs, resulting in a number of hospital visits. With no downstairs bathroom and limited space to manoeuvre there were a number of accidents and breakages. Ben had argued that the support network from their neighbours was very important, particularly in a crisis, as they had a school-age daughter. However, the council was able to find them suitable accommodation nearby and once the family had settled they liked their new home.

Sophie and Melanie had previously lived in London. As Sophie's condition progressed the opportunity arose for them to buy the bungalow next door to where their mother lived and cared for their sister, who also suffered with HD. More recently as Sophie's condition deteriorated further Melanie paid for a wet room to be installed to make washing easier. Julia, Martin's wife also incorporated a wet room into the renovations funded by Social Services to create living spaces for Martin in the downstairs areas of their house. This allowed Martin to have access to everything he needed within a short distance.

Well we're having alterations done to the house. We've got a wet room, we've got a bedroom. Through there is the bedroom and a wet room. But what I want is access for Martin to go straight from the bedroom into the dining room and the kitchen on the far side and have it as a dining area through a door ... Access all around so then it'll be a bit like a bungalow for him. ... I've tried to think ahead as much as I can as regards getting things done. (Julia, Martin's wife, Interview 1, 12.02.2008)

It took some time for Julia to get all the adaptations made to the house as requests were often stalled by what families considered to be bureaucratic processes and an unnecessary division of labour. I returned to interview Martin and Julia for a third time in August 2010, over two years after she had initially requested the door, and the building work had begun on Martin's downstairs bedroom. In order to get an additional door between the kitchen and Martin's bedroom it took several requests from Julia as she was initially only allowed a window as she describes:

I was told that couldn't be a door. ... So when they did the alterations they put a window in, and I let it go for 18 months, and I reapplied and got the assessment again. And suddenly it was legislated that I could have door as long it was a fire [door]. ... And it goes a bit like that, not being funny, I am very grateful for everything I have, but god is it a fight (whispers). It doesn't matter what it is, and it is not the people, it is the way the, it is all sorted out, there are that many departments and they can't do it but they

can, and they can do this, but they can't. (Julia, Martin's wife, Interview 3, 18.08.2010)

After several months living at home Vicky was placed in a specially adapted flat, provided for young people wanting independent living. It had a lift, widened corridors and an adapted bathroom. Regaining this independence was vitally important to Vicky and despite the fear of letting her daughter go, Vicky's mum, Sheila recognised how essential the move had been for both of them.

I did think that when I was at home, before living here by myself, that was the worst time. Mum has got an upstairs and I used to find it really difficult going upstairs, so I used to just live downstairs in the living room and I didn't go up and have a shower, I didn't have bath, I didn't do anything. I just lay there, all day, all night. ... (Vicky, Interview 2, 30.09.2010)

From seeing her lying on that settee, day in and day out, not even having a bath, not changing her underwear, not getting up and been grumpy, ... her not going upstairs to bed, so sleeping there, eating there, lying there, it was just horrible. And it was a repeat of [my husband] doing exactly the same, and it was just like all of my sort of space in this house was confined to this room, didn't even want to go down that area because that all was Huntington's. (Sheila, Vicky's mum, Interview 1, 03.08.2009) ... I don't think either of us would have had the quality of life if Vicky hadn't moved out. (Sheila, Vicky's mum, Interview 2, 19.10.2010)

Adaptations were designed to maintain independence for as long as possible and make it easier for someone to be cared for at home when they could no longer physically manage personal care, such as washing, dressing and getting to bed. As these demands on the family carer grew additional help was required.

It was not just alterations that changed people's home into a hospital at home. Families often had to store large amounts of resources such as PEG feeds, incontinence pads, nutritional supplements, and find place for equipment including walkers, wheelchairs, specialist beds, hoists and commodes. To add to this invasion of the home space numerous health and social care professionals

also visited patients and families at home. At this stage in the trajectory of the condition it was up to family carers to manage all these services but families reported that it was often difficult to know who all the health and social care professionals were and why they were there. By the time I conducted my final interview with Julia in April 2011, Martin was no longer able to take part and Julia explained the various people they had coming to the house. These were in addition to the home carers who came three times a day and stayed overnight.

Julia: Well I've got my dieticians and all that sort of thing now, as usual I've got [speech and language therapist, the CNS, the consultant], we keep all them. His GP, nutrition nurse now he's being PEG fed ... Oh incontinency nurse, have I said her? ... trying to think what else we have, I think that's it. I have to write them all down, they're all in a book. I just get the appropriate one when required.

Interviewer: You have a lot of people coming and going don't you?

Julia: Oh this house never stops, they'll be all in again, one o'clock it will start off, and then they'll go at two, and then they'll be back at four, then they'll go, then they'll be back about half past six, seven o'clock. They'll finally go at eight o'clock, then the carer comes at half past nine, 10 o'clock, the night shift. She actually starts work at 10 though. And it all kicks off in a morning, I get up half past six in the morning to get Martin shaved, fed before they come in. (Julia, Martin's wife, Interview 4, 19.04.2011)

Ben also talked of trying to keep track of the number of health and social care professionals associated with Amy's care. When we tried to complete Amy's eco-maps the couple were often not clear on how many, who the health and social care professional were, or the reason for their involvement. They were familiar with a core team including the consultant, the RCA, the home care service who visited twice a day. However over the study period they also reported input from a number of different social workers, occupational therapists, a one-off referral to the Birchtree HD team, as well as physiotherapy, speech and language therapy and hydrotherapy services. This myriad of services often left them confused about who was contacting them and why as well as whom they should contact for their various needs.

SERVICE INPUT

During this mid phase people with HD and their families encountered a myriad of services. As their needs increased services needed to be put in place to allow the person with HD to continue to be cared for at home. The patients in this chapter had a range of 'care packages' made up of home care services, such as washing and dressing in the mornings and evenings; day care services, where the person with HD would be picked up and taken to a community centre for the day to engage with other people and be entertained with arts and crafts and so on; and respite, each family was allocated a number of weeks per year when the person with HD could go to a residential care centre for 24 hour care, allowing the family carer a break from their caring role. Sitters were also used by some families to provide cover when they wanted to go out in the day yet could not leave the person with HD alone. As a young person in her early 20s, one participant, Vicky, had a personal assistant. This person was to help her maintain her social and physical activities and would assist her in going where she wished to go during the day such as to the cinema, swimming or shopping.

In 2010 and 2011, towards the end of the study, the economic downturn meant that cuts were made to health and social care services across England.

Participants noted, in particular, greater difficulties to accessing day care and respite services. This change in economic climate is reflected in families' stories about the services they received and the challenges faced in accessing these. It is also evident in the reports of health and social care professionals who felt the burden of further financial constraints during this time.

DAY CARE

Day care services varied across the different geographic areas and were often provide by local councils or charitable organisations and included transportation services to and from the hosting centre. Day care services were often introduced as first option to encourage independence for the person with HD and their family carer in having some time apart. They were also seen as a way to keep the person with HD engaged in hobbies and social activities.

Participants consistently reported enjoying attending day care services and found this a valuable resource for both people with HD and their family carers.

He does go to day centre three times a week, which is very helpful. But it's good for Martin as well to socialise. (Julia, Martin's wife, Interview 2, 15.12.2009)

Amy invited me to attend her day care service with her to see what she did during the day and to show me the wooden rocking chair she was making. The centre is funded by a charity for people with a range of neurological disabilities and is supplemented by a £3 fee for the day. Amy is picked up at 9am by the transport provided and dropped off at home at 3.30pm. The centre was based in a large council building over the majority of the ground floor.

We enter to a reception area and Ben introduces me to staff. There are some small rooms off the corridor - one was being used by a woman and a carer helping her do some word puzzles, one was an aromatherapy room where they go for relaxation and often a nap in the afternoon - the woman who does the sessions explained how she does Reiki (relaxation massage) with Amy and all her movements stop, so they are going to get the CDs to see if it will work at home. There is also a woodwork room, which seemed to be the hub of the place with four people in the process of making things. Amy is making a rocking chair with the help of a woodwork teacher and some carers who seem to just get involved as and when needed (apparently they are mostly allocated to one person but all chip in). Opposite the woodwork room is a dining room. We took a morning break there for a cup of coffee. (notes on observation 2, visit to Amy's day care centre, 29.06.2010)

For Sophie and her sister (also affected by HD), day care was a time when they could spend time together without their family carers. It also gave Melanie and her mother some free time together. Again funding of these services was an issue and was more acutely visible for Melanie as her sisters were allocated different funding allowances because Sophie had a pension and her other sister did not. Melanie therefore had to weigh up the advantages of Sophie attending

day care and the cost of that care.

Sophie goes now, she goes four hours on a Saturday and four hours on a Monday. ... you've got three hours or three and a half hours that you could actually go and do something which is quite nice. So that's good, but it's all into the money though, I mean that's the problem. I think it's quite reasonable, they only charge, they do charge £6 an hour which is more of a realistic amount to pay. But it's still at the end of four weeks you're looking at £200 which when, for someone, especially now I'm not working it's quite a big chunk of [money]. I weigh it up, whether it's worth [it], and it is. For her and for us it's a good investment. (Melanie, Sophie's sister, Interview 4, 02.06.2011)

HOME CARE

Prior to her husband's stroke Sarah had a care package in place incorporating home care both in the mornings and evenings. Max talked about the importance of this home care service in supporting Sarah to remain at home. This sentiment was echoed by Ben as he recognised that he would not be able to cope with Amy's care at home without the home carers coming both in the morning and at night to tend to her personal care.

I now have a package of help from the local authority. And they come in in the morning to get her up, just one girl comes in in the mornings, it's easy in the mornings. And then at night two girls come in to shower her and everything. ... and they put her to bed. So it works. (Max, Sarah's husband, Interview 1, 27.11.2007)

The need for home care services was often brought to the fore for family carers when they recognised that the person with HD could no longer be left alone for any period of time. This prevented them undertaking even the smallest tasks in the day without assistance. Martin and Julia used a sitting service to allow Julia time to attend her own doctors' appointments, go to church and visit their daughter. Max recognised his social time was curtailed when he could no longer leave Sarah in the evenings and he needed to adjust his routine to accommodate

all his household tasks, such as shopping, when Sarah was at day care. For Ben an additional midday visit from home care services meant that he was able to run errands.

Ben: Amy likes to go out but she's not well enough sort of thing, so sometimes she's stuck here so they organised for a carer to come in about one o'clock in case I've had to nip and run some errands or do a bit of shopping or whatever, you know, just to make sure that she's got everything and she's okay and not in a big heap on the floor...

Interviewer: What do you think would happen if you weren't able to have carers in?

Ben: I wouldn't be able to cope. I would not. I'll admit it I would not. If I went shopping I'd have to take her with me. Wherever I went she'd have [to come], I wouldn't be able to go out without her. (Ben, Amy's husband, Interview 3, 10.12.2010)

Sophie had been on the cusp of this mid stage for some time. Sophie and Melanie had managed without needing services for Sophie's personal care for nearly 20 years. Sophie had started to access day care services over the past few years and was now becoming increasingly dependent putting increased pressure on her sister to be available 24 hours a day and help with personal care. In their latter interviews Melanie expressed the strain placed on her now that her sister could no longer be left on her own.

She was still working but I'd taken, it's been 10 years, no 12 years since she stopped working and so since then it's been sort of a full on. And I think I've just noticed it more because so many things have been taken away from us this year, and she has deteriorated to the point where I can't leave her on her own for, it's probably all right for half an hour if I literally have to pop to the Post Office ... And that's quite hard, that just curtails anything that you want to do spontaneously, you can't do anything spontaneous, I'd say it is real major planning, I think that's what frustrates me sometimes. (Melanie, Sophie's sister, Interview 4, 02.06.2011)

RESPIRE

Family carers considered regular respite care to be an essential part of their support network. It was the brief period when they could take a break from the emotional and physical labour of caring and take some time for themselves. As Julia explains

It's always very beneficial, because it's a total change from routine and it's just mentally resting. ...Yeah, it's always good to get right away because if I stay I still get the phone calls, I still get everything, you know. But I am ready, once that two months is up, you can tell, I'm very edgy, I'm ready for the rest. (Julia, Martin's wife, Interview 2, 15.12.2009)

During the early part of the study participants talked of booking up their allocated respite periods. The system requires respite to be means tested so most were allocated some periods free and then had to a pay contribution towards any other weeks they used. However in more recent years there have been spending cuts which have created uncertainty for families as to whether they will continue to receive funded respite periods and whether these allocation will be considerably fewer than they had previously been. Family carers were dependent on respite periods for a break from their caring role. Like Julia and Melanie they recognised that after long periods of caring they became tired, irritable, stressed and exacerbated their own illnesses.

...hearing on the news about their not having a respite packet, they're taking all the respite away. People are going to be angry. ... But I think we're going to have more and more problems with people who physically, you can only go on so long without, I found myself yesterday shouting at her for no real reason at all, just that she was irritating me. But she wasn't, I mean she was just being Sophie and if it hadn't been my sister I wouldn't have got angry with her. Her speech is deteriorating quite badly, I think I get frustrated that I can't [understand her]. (Melanie, Sophie's sister, Interview 3, 21.01.2011)

Another element of uncertainty was created when the residential care home used by a number of participants and others with HD in the Birchtree area closed its respite beds. This forced families to look for other places that would accept clients with HD. However, for people with HD, particularly at this more advanced stage of cognitive decline, changes in routine or familiar faces could be an additional stressor.

Sophie's devastated because I really sort of let it slip only a couple of days ago because I just hadn't mentioned it because she knows she goes to respite but I hadn't mentioned that [the unit had closed], I wanted to only let her know when I'd found somewhere else ... of course with Huntington's they like the routine. Even though it may not have been the best, but to them it was great because they knew people there and the people knew them. (Melanie, Sophie's sister, Interview 3, 21.01.2011)

Towards the end of the study period Melanie explained the importance of their allocated time for respite. She had serious concerns about the number of respite weeks being reduced due to recent government cuts on spending in the current financial climate. However it was not clear at this time how this would affect their family directly, leaving Melanie with considerable uncertainty. With little other support Melanie regarded these breaks as vital to her ability to continue to care for her sister. She felt that to date she had only been able to cope because she and her mother worked together to share the caring roles necessary to look after both her sisters.

Up until this year we've had four [weeks] a year, and whilst in some respects it hasn't been enough it's certainly been nice knowing [we have it]. At the moment I don't know if this one week that Sophie has in July, if that's the only one I'm going to have. ... 11 hours a week I don't have Sophie. ... That will be the one thing that I think will cause a lot of people to finally give up, knowing that they haven't got any more respite, there's no respite weeks. That's the thing for me ...that will cause more stress than anything else. ... And it's as much for Sophie as it is for us, so that's my major

concern, I just don't know and nobody seems to be able to tell us. (Melanie, Sophie's sister, Interview 4, 02.06.2011)

RESISTING SERVICES

Not all participants welcomed the increasing levels of input from services. Amy was particularly resistant to any services outside of her home. She had home carers and attended day care two mornings a week. For Amy this was the only time she was separated from her husband as she had refused respite care. Despite enjoying her time at day care she would only agree to go for two mornings a week. This particular family had complex and often strained relationships with a number of services with which they had engaged. With a young daughter living with them, Social Services also maintained contact with the family. Ben's ability to provide suitable care for their daughter, Isabelle, and Amy was questioned a number of times. For Ben, Amy's few hours at day care two mornings a week was the only break he got from his caring role. He recognised he was struggling to continue without a rest and mentioned this to me several times during informal conversations at clinic and when Amy was not nearby. However it was difficult to discuss this with him further as I was never able to interview him on his own.

Well she only does half a day at the moment, two [full] days is too much. ... she's getting too tired on a full day there. (Ben, Amy's husband, Interview 3, 10.12.10)

I had arranged an interview while Amy was at day care but because of adverse winter weather her transport was not able to run that day. Amy's bed was in the living room of the house and there was nowhere else to go to have a more private conversation. However while visiting Amy's day care centre with the couple I was able to have an informal conversation with Ben.

Amy goes two days a week and really enjoys it but doesn't want to go more. Ben said he was getting to the 'end of his tether' and was really struggling to cope now. Amy refuses to go to respite even though when she visited [a local unit] she loved it and wanted to stay there and then

but once given time to think about it she changed her mind and then they missed their opportunity as spaces are so scarce. Ben said the RCA is looking into getting carers in so he can go away as really needs a break. (notes on informal conversation with Ben during an observation at the day care centre with Amy and Ben, 29.06.2010)

Despite the efforts of the RCA and consultant Amy continued to refuse to leave her home for respite care, concerned that once she was there her husband would not take her home again. During their third interview Ben and I did talk a little about respite in the context of exploring a new in-patient drug regime for her extremely pronounced chorea. However, he explained that even to go to hospital for this symptom control Amy wanted a contract to be signed to say she would only be there for a set period.

There's this contract thing got to be drawn up that she wants. ... basically saying that she'll go and it's for X amount of time, and it's X amount of time only. ...Because she's got it in her head that everybody's trying to put her in ...a [care] home ... (Ben, Amy's husband, Interview 3, 10.12.2010)

Amy's fear of being left in a care home was persistent, but in my notes I queried whether she was able to understand that by not allowing Ben some time to rest she may have been curtailing her chances of being able to remain at home in the longer term.

TAILOR MADE

There was an attempt to make care packages tailored, not only to the health needs of the individual but to the individual themselves. Providing this level of service to people poses a number of challenges, not only in terms of both the rarity and complexity of HD as a condition but also the range of people affected by the condition. As Vicky shows, it has not always been possible to get services to meet her unique needs of being a young person with a number of evident symptoms of HD. Vicky's home carers help with personal care tasks as well as

activities of daily living. In addition she has a personal assistant [PA] to help her maintain as many of the physical and social aspects of her life as possible.

Vicky: They come in the morning for two hours and help me shower and get changed. And they come at lunch time to help me eat my food and then at dinner again for my food.

Interviewer: Do you have the same carers every day?

Vicky: Yeah

Interviewer: Do you get on with them?

Vicky: Yeah yeah they are nice. ...Yeah because I did have, before [this PA], I had a different PA lady ... because she didn't drive, that didn't help, because I couldn't really go and do stuff. She just would come and like make some food for me and then she had to go back again.

Interviewer: So has [your personal assistant] worked with people with Huntington's before?

Vicky: No, she worked with somebody who had got Down's Syndrome.

Interviewer: So is she learning from you then?

Vicky: Yeah yeah. I think most people do. It's not very common. A bit like the carers who come in the mornings, at first they didn't really know because the people that they work with are just elderly people so I am the only young person. (Vicky, Interview 1, 17.12.2008)

Martin struggled to come to terms with his loss of independence and the thought of a stranger tending to his personal care. He had resisted home care for as long as possible, but was aware that his wife could not manage his needs. The care agency and sitting service were able to provide male carers, this helped Martin to feel more comfortable and form easier relationships with sitters when they came to keep him company.

Interviewer: So what do they do in the morning?

Martin: Shower and dress me, have a joke.

Julia: It's the men's talk. ... [the home carer] is good for him. Martin's never been the same since his carer's been coming here. It's all men's talk in there

isn't it in the morning, oh yeah. You like [the home carer] though. (Martin and wife Julia, Interview 1, 12.02.2008)

As Julia's own health was considerably impaired with both angina and diabetes Martin's care package was tailored to accommodate Julia's disabilities as well as Martin's care needs. Martin also suffered with a sleep condition which caused continuous disturbances in the night. In order for Julia to care for Martin in the day she needed to sleep at night, so night sitters were also introduced. The couple also had domestic help in the form of cleaning and ironing.

DEMANDS OF THE DISEASE

All participants reported home carers having little if any experience with HD and having to teach them to manage different aspects of the condition. This highlighted the importance of continuity of care across all the services. The participants with HD struggled to cope with changes in carers or routine and this could heighten behavioural issues. For family carers changes in home care staff meant the additional effort of having to teach a new person about the individual and their particular abilities and behaviours.

He's supposed to have a team of carers, the same ones, because of his condition. Oh no, they've been sending different ones. So I've been in touch and complained, I said look you promised me, which they did, they promised me a team of four to stay with Martin for Martin's care. ... I'll PEG feed him if I have to but I mostly let them do that. Because none of them knew how to PEG feed. I had to show them all ... So it was a good job I was shown, so I sort of taught them all how to do the PEG feed one by one as they come in. (Julia, Martin's wife, Interview 4, 19.04.2011)

HD often causes behavioural issues and it was apparent from the stories of family carers that these issues were challenging for home care staff. Max reported a number of behavioural issues which manifested as part of Sarah's condition. However most disturbing was her tendency to lash out at people she did not like, and she could behave violently when she was asked to do something she did not want to do or was out of her normal routine.

Sarah didn't like the carers, some of them smoked so she could smell the smoke in their clothes, they wore jeans and she can't stand jeans, and she used to tell them. And one girl she took a particular dislike to ... if Jess came, oh let's hope there's a good night tonight, and there rarely was when Jess was here. I mean one of the evenings in the bedroom she took a real dislike to her, and she just rammed her and pinned her against the wall and sort of threatened her, her arm was raised and everything. And if the girls came before 7 o'clock, she used to watch the news between 6 and 7 every night, if they came before 7 o'clock then it was hell, because she was totally non cooperative, she wouldn't do anything until the news had finished. (Max, Sarah's husband, Interview 2, 12.03.2008)

One of the most problematic behavioural traits Vicky had developed was a combination of fixed thinking and a lack of impulse control. This resulted in Vicky needing her wishes to be met almost instantaneously. This was extremely demanding for her mother and caused problems for Vicky's Personal Assistant when her repetitious demands continue beyond working hours.

And one of the carers actually was in tears, she's [Vicky's] age and she said to me, she said I don't know how to handle it, she keeps on texting me and I've got a baby and ten o'clock at night she's texting me about whether I've remembered that we're going somewhere tomorrow. ... [I said] you need to sit down with her and just be honest with her say look you're off duty when you leave here, yes in an emergency you've said that your phone will be on for you, for her if she can't get hold of me which is very kind of her. So I did speak to [the Personal Assistant] and I said 'well what did she say?' She said oh, she said 'I know I do it but I can't help it', which is even worse really, it's like you feel, partly I thought well if she doesn't know she does it then it's like, it's easier, but she knows she does. (Sheila, Vicky's mother, Interview 2, 19.10.2010)

BALANCING RISK

Throughout this period both family carers and health professionals grappled with balancing the safety of the person with HD and allowing them the freedom

to live their lives as they wished. Key areas of risk noted by family carers and health professionals were eating/choking and walking/falling.

The speech and language therapist was working with Martin and Julia to educate them on the risks of choking and teaching them techniques to avoid choking episodes.

I have been involved with him in terms of his swallow. With him it's just monitoring, making sure he's on the right consistencies, and also educating his wife in order to give him the right consistencies [of food]. I'm very aware that the carers are with them 24/7 and they do know them better. But sometimes they do not appreciate risks of what could happen if they give them certain consistencies. With Martin and Julia it took a while for Julia to realise she was putting him at big risks with the consistencies that he was having. The first time I went and watched him she'd given him some sort of a fish pie and he really did choke badly. ... So I had to talk to her- and I would sit with her, I have brought diagrams about where the food is going and what is happening. (Speech and Language Therapist, Interview 1, 21.09.2010)

The speech and language therapist had also been involved in educating the staff at the day care centre Martin attended to make sure he was also getting appropriate food when he was there. At this time there was little the speech and language therapist felt she could do to improve or maintain Martin's communication so her focus remained on sustaining his swallow and eating for as long as possible.

Because we can't physically just keep ringing up just to check if they are ok and try to educate the carers and key worker [at the care centre] to take the responsibility to contact me. I don't think we have got the capacity in the NHS to ring them regularly. And Julia is pretty good at ringing me if she thinks there is a problem. I would either go out and see him or talk it through on the phone. ... It's just making sure they can manage as best they can for as long as they can. (Speech and Language Therapist, Interview 1, 21.09.2010)

Despite her concern about Martin's swallow the speech and language therapist also felt the timing of having a PEG was important. She wanted to balance the elements of giving up eating orally, including its social aspects with a decline in his physical condition. Having a PEG fitted is a relatively minor operation but does require the person to be in sufficiently good health to cope with this intrusion.

But obviously we don't want it to become a crisis.... So it's trying to get that right. ...I think if we go down the route of a PEG too early, even though that's what he says he wants, he will stop eating too soon. I think that would be a shame. He enjoys his food. He enjoys sitting in the dining room at the [day care centre], and seeing everybody. ...It's such a sociable thing-eating. (Speech and Language Therapist, Interview 1, 21.09.2010)

As Martin's swallow deteriorated his incidents of choking became more frequent. He had also had several bouts of pneumonia, although it was not clear if this was caused by him aspirating, his asthma or his increasing lack of mobility. In late 2010 the idea of him having a PEG was reintroduced. In March 2009 I had observed Martin's yearly clinic visit. In discussion with the CNS prior to the start of clinic she told me that as this was Martin's yearly check-up they would revisit his advance directive which related to his decision to have a PEG when necessary. This was done in his consultation with the consultant and the CNS present and Martin reiterated his wishes to have a PEG when he could no longer eat. It was also raised in his consultation with the speech and language therapist and dietician. The dietician took some time to explain the process to Martin and show him an example of the PEG tube so he could see its size. This extract is from my observation notes during this clinic appointment and Martin's visit with the dietician.

Again [the dietician] is aware of the Advance Directive for the PEG. ... She goes through the leaflet on PEG feeding with them: Explains it is a tube that goes into the stomach but you can still eat and drink as normal if you wish. The dietician explains it can be used to give some medications if necessary. Martin asks if you can feel it when it is in place. She explains it

just becomes part of you so you don't feel it. (notes on observation of Martin's clinic appointment with the dietician, 25.03.2009)

The decision to have a PEG had been raised with Martin, and by Martin himself, several times so when the final decision was made to have it put in Martin was in full agreement. The PEG was finally fitted in March 2011 after several delays due to Martin's ill health.

Fear of injury from falling was another area of concern for both family carers and health professionals, although little anxiety was displayed by those with HD. People with HD were encouraged to walk, even for short distances, to maintain muscle strength, and the benefits of different bodily positions. Many participants reported falling, and mostly only incurred minor injuries. However, as people became increasingly unsteady on their feet and the number of falls increased concerns were raised about their safety. Amy reported a number of falls over the study period and this was one of the key reasons for their move to a single level adapted house. Max also reported a dangerous incident when Sarah smashed a glass door as she fell. Sophie was still ambulant over short distances and in the home so it was only with her more recent deterioration that Melanie began to fear her falling if left alone.

Interview: So what's your concern with leaving her?

Melanie: Falling. Falling or she would try and boil the kettle. ... I mean the difference being if she falls and I'm here, I'm here and we can somehow get her up but if she fell two minutes after you've gone out and you're not back for two hours it's a long time. As it is she would probably get herself up, she probably would cope okay. I think it's more my worry more than hers, it doesn't bother her at all. (Melanie, Sophie's sister, Interview 3, 21.01.2011)

Living independently Vicky had more control over the services she interacted with. However this caused concern for her mother when Vicky sent her carers away without help to get up, washed or dressed. This lethargy and lack of motivation is common in people with HD but can result in some behaviours that might put their health at risk. Although concerned about Vicky not washing and dressing Sheila's fears were for her wider safety in relation to swallowing and

falling. As a mother and carer she grapples with balancing respect for her daughter wishes and concerns for her comfort and safety.

...and especially, she's not coming out of bed to eat now, she won't eat at the table which is not good either, for her choking it's not good. She's becoming quite a recluse into her bedroom and she won't let anybody touch it, it's really messy but she won't really let anybody clear up, so that's hard. I try to do it but she wants all her things around her in the way she wants them, which is fair enough really, but it's clutter and I'm scared that she's going to trip over something. And I think it's just that, how much do you intervene and how much do you, and it's so hard, it really is, it is hard. ... All the time it's about sort of the freedom of my daughter and wanting to get the best for her and it's like how much do I intervene, how much do I go behind her back, how much do I sit down with her, and that takes so much energy trying to get it right and I never get it right. (Sheila, Vicky's mum, Interview 3, 14.07.11)

This concern was not only for Vicky's physical safety. As her daughter became increasingly cognitively impaired and her motor skills declined she struggled to maintain additional activities of daily living such as managing her finances. Sheila's concern therefore, was that her daughter be protected from potential mistakes, or someone taking advantage of her vulnerability.

[Vicky] keeps on going on about the power of attorney. I haven't even raised it, it's her that keeps on saying 'oh mum will you sign my cheques'. And usually she's been very protective of all her paperwork. ... I think she's paid through the online banking, three or four times to the same person, and I've just picked that up from her statements, so she's pressing the button and not realising, so people have been getting overpayments and then it's that trying to chase that money and trying to get it back, and it's stuff, so... But it's about trying to do it with dignity so that she's still in control of her life, as much as she can be, but also to make sure that she's not being abused really. (Sheila, Vicky's mum, Interview 3, 14.07.2011)

Supporting Vicky is particularly challenging for Sheila as she continues to battle with her mother-in-law in India about the secrecy and silence that surrounds HD in their family. Vicky's grandmother refuses to acknowledge HD in the family despite her husband and son dying from the condition, and her daughter and granddaughter being diagnosed and symptomatic.

FUNDING AND BUREAUCRACY

Each of the participants in this phase were eligible for state assistance through both health and social care services. However, as Sophie was only more recently entering the phase where she required help at home means testing prevented her for accessing the same level of services her sister had been provided with. Sophie had worked as a teacher and therefore had a pension. At this stage in her illness she did not warrant funding for home care. Melanie received a carers allowance, however this did not cover home care provision so any additional services would have to be paid for by her pension. This was a source of frustration for Melanie, who felt the discrepancy between the health and social care provision for her sister living with her mother and Sophie was unfair.

Technically Sophie gets an allowance for [home care] obviously in her Incapacity Benefit but because she gets a pension it would cost us more than her allowance to have somebody here for 40 minutes a day to get her up in the morning. I think it works out around the £400 a month ... they give me £53.90 a week to look after Sophie for a minimum of 35 hours a week which is something like a pound an hour. Realistically I look after her 24 hours and she's never on her own so she's cared for 24 hours 7 days a week ... £54 really to look after somebody yet if I had a carer in they would charge me double that for 40 minutes a day. (Melanie, Sophie's sister, Interview 3, 21.01.2011)

However it was becoming apparent that Sophie's condition was deteriorating and at the end of the study period she needed home care support. The CNS for Birchtree involved in overseeing Sophie's care also recognised the importance

of respite to maintaining Melanie's coping mechanisms and therefore maintaining care for Sophie at home. The CNS reported on the deterioration in Sophie's condition and her need for additional input, particularly in the last year of the study (2010/2011). Sophie's speech, balance, swallow and cognition had all deteriorated and the CNS noted the challenge of getting commissioners to recognise the physical and cognitive effects of HD.

The OT has been in now I think and we are looking at doing another continuing health care needs assessment. Because I don't think Health [funding bodies] are recognising the cognitive decline in her and that could enable us then to click in to get more funding which will then support her with either home care or respite care. I think that is imperative in maintaining her at home, and for maintaining the carers health she needs regular respite, and regular home care. Someone either coming to take her out or sit with her whilst her sister can go out and do things. (CNS, Interview 3, 27.06.2011)

Funding and bureaucratic processes caused a number of problems not only for families but for the health professionals working with them to provide the most appropriate care. Gaining access to services was always considered complex and challenging for families but recent cuts in service funding meant that care packages were being reviewed. In both Birchtree and Oakfield the key health professionals such as the CNS and the consultant contributed to all assessments and reviews of care packages in order to provide the correct information and clarify understanding of the condition and the complex needs it created. As funding for care packages could be provided by health care, social care, or a combination of the two it was important for those making assessments to understand the needs of those with HD. However this was not always the case as Ben reported.

Social Services are doing what the hell they want as normal. ... she doesn't meet the criteria for care anymore. Well [the consultant] called it a joke. ... [the consultant is] doing me a letter that I've got to sign because they're not listening to him. So he's writing a letter as if I've wrote it, because he's

hoping then they'll listen to me as carer, because he wants that meeting held again where he can be there and [the RCA] can be there. (Ben, Amy's husband, Interview 3, 10.12.2010)

Those health care professionals working closely with families who had seen the impacts of the condition on the person with HD, the family carer, the wider family and even the home itself were often frustrated by those making decisions about funding for care provision. The CNS for Birchtree had known Martin and his wife for a number of years and understood the complexity of his condition alongside that of this wife's health problems. Here she explains how and why she wanted to increase his care package to include night sitters.

They questioned, for instance, as you know his wife has got her own health problems. He's also got a sleep disorder, which means that he's very active even when he's asleep. ... One of the issues about supporting carers is they need to have their rest, ... Julia understood that she's got to get her sleep because she can't manage him during the day. She gets touchy with him and it can just break down quite easily at home. So one of the things I'd asked for a sleep-in service seven nights a week. That was knocked back to four nights. The excuse being well he actually goes to day care for three days so she can sleep those three days. It's things like that that I don't know how they can make that judgement when we've actually asked for something. But they've made the judgement that when he goes to day care for three days, so those nights before he goes to day care she doesn't really need to sleep because she can sleep the next day. (CNS, Interview 1, 24.06.2009)

As resources are scarce carers often felt they had to fight for the allocation of services. This was coupled with a generally limited understanding of HD by service providers. It was often considered to be a mental or physical condition rather than encompassing both aspects of the disease. These conflicts and ongoing fights for equipment and services placed additional strain on already pushed family carers to deal with bureaucratic processes as well as a relative with complex and demanding needs.

You'll find you're the co-ordinator, if you're the carer you're the co-ordinator, or you get forgotten. ... If I get really stuck I ring [the CNS] and then she'll come in, well she knows more people than me. ... Because between looking after them, you're ringing one for this and one for that, complaining about the other, and it is a bit, can be a bit, and if you can't get the answers to what you want it can be very stressful, very very stressful. (Julia, Martin's wife, Interview 4, 19.04.2011)

Key health care professionals like the CNS, the RCA and the consultants acted as advocates and liaisons between services. They were also considered to be the first port of call in a crisis, emergency or problem. As Julia describes the CNS had been part of Martin's care since his diagnosis in 2003 and although she felt she now could manage most things she still considered the CNS an essential part of their lives. She relied on her for support and to plead Martin's case when she could not.

Julia: The only person that has really backed me has been [the CNS], she has always been the one. She is the one that encouraged me to go for [the alterations to the house], worth her weight in gold that one. ...

Interviewer: So it is helpful having her, somebody dedicated for HD then?

Julia: Oh oh yeah. Oh well you haven't got a chance otherwise to be honest. ... We have a lot to thank [the CNS] for, in those early days, very very much. Well still, but we don't need, we have got used to it now. Our [care] package is in place. Even the funding, which really isn't [the CNS's role], it is not down to her, it is her going that extra mile for us. She has been marvellous for all the Huntington's people, not just one. ... if you are stuck and you are worried it is [the CNS] we all head for. ... I don't know what we would do without her, really I don't. (Julia, Martin's wife, Interview 3, 18.08.2010).

The CNS saw her role as one of support and advocacy in these types of instances. The longevity of the condition meant that rapport and trust could be built over time. However knowledge of the condition and the other types of service available was vital to direct families and advocate on their behalf.

LIFE BEYOND 'CARING'

All the family carer participants talked of the challenges of being a 'carer' and providing care to their family member. Melanie and Sheila in particular noted the different challenges they faced by being a sister and a mother of the affected person. All the other family carers were spousal. Melanie and Sheila also had other close family members whose care they had been or were involved with. Melanie also helped to care for another sister who lived with their elderly mother. Sheila had previously cared for her husband with the condition. She expressed the distress of slowly losing her daughter to the same disease.

I think it's just seeing Vicky's deterioration, it's just too painful really, ... because I'm down, I'm on a short fuse and I'm not as tolerant of Vicky, and it's not because I'm not tolerant of Vicky it's the behaviour that, it's like a reminder of everything, of [my husband] and everything, it all just brings it back. So poor Vicky gets the brunt of that, because it's not about my encounter with her, it's everything else that's happened as well, and the future that I see, so every encounter with her is about all of that in that split second. ... you can't help it because that's who we are and what we are really. I try not to and then I blame myself, and I think, well actually it is what it is and make the most of whatever we have got. It is about living in the moment, trying to enjoy what we have. (Sheila, Vicky's mum, Interview 5, 14.07.2011)

As she was not a spousal carer Melanie reported being challenged by people as to why she undertook providing such extensive care for her sister.

It's no good someone saying well you don't have to take care of them. I think no I don't, if it was that black and white no I don't have to take care of Sophie but if I don't take care of Sophie she will have to go into full time care and she doesn't want that. ... I wouldn't want to either. So it's no good someone saying to me you don't have to do it. I said yes but I have to live with my choices and that's huge. I mean if I couldn't take care, that's the difference, if you physically can't do it or you don't have the space or

*anything like that then fine but when you can do it it's a big thing.
(Melanie, Sophie's sister, Interview 3 21.01.2011)*

For family carers trying to balance their own life alongside their increasingly demanding role as a carer became an additional challenge. Their time was increasingly taken up by caring tasks and once the person with HD had reached the stage where they could not be left alone, free time, and time to complete errands, or engage in social activities, was progressively encroached upon. Julia used the sitting service to allow her to continue to attend church and take some time to visit her daughter. Ben slowly gave up his activities outside of the home but took advantage of the times when the home care services were tending to Amy's personal care needs to take a break and relax with some video games.

For family carers like Max it became necessary to give up work. Melanie also struggled to manage work and her caring role. However when she was made redundant in early 2011 she became concerned with re-establishing her place in society. As a person in her 40s she felt it was important for her to contribute to society and she valued the role having a job afforded her. A reduced income was also a considerable worry but Melanie's concerns were around having something for herself as well as contributing to the household.

*I was made redundant at the beginning of December which has a mixed blessing. The job I did was great because I could work at midnight and 5am, it didn't matter what time I worked ... but financially so you look at it in a different way. ... I have the guilt factor I'm not working, but I am. I take care of two people full time but I've never not worked. Since I left university I have never not had a job and I suppose there's a part of me that might resent it slightly because I can't go and do what I really want to do ...
(Melanie, Sophie's sister, Interview 3 21.01.2011).*

Aware that this was a long-term condition meant that life could not be put on hold, family carers tried as far as possible to continue to undertake ordinary activities such as home improvements and going out for the day. These ordinary tasks were supplemented by activities constructed to 'make the most' of the time they had while the person with HD was well enough. These included

holidays and special trips. Sheila describes a number of special trips organised throughout the three years she and Vicky participated in the study. Each of these were part of fulfilling as many of her daughter's wishes whilst it was still possible to do so. With the help of a friend she was able to take Vicky to Australia and the whole family took a trip to India for a family wedding. She also arranged smaller excursions such as weekends away to allow Vicky to spend time with her nephew.

It's about trying to fulfil any of Vicky's dreams and all of Vicky's dreams that are within my capacity to be able to do really. And it's not just about coasting on and dying, it's about actually making the most of life really, and knowing that it's going to be more enjoyable when she's as mobile as she can be really. ... I remember when her nephew was born she said I'm not going to love him, I'm not going to be here when he gets older so I'm not going to get attached to him. ... [but he adores her and] he is not hers but she's trying to sort of get that enjoyment really. (Sheila, Vicky's mother, Interview 4, 19.10 2010)

Melanie also arranged a number of trips for her sisters in order to make the most of the time that they were still ambulatory enough to participate and enjoy these experiences. These were often local outings such as to the theatre, cinema or church group, but like Vicky were sometimes bigger trips abroad to fulfil wishes. Family carers noted that they tried to balance not planning too far ahead with continuing to participate in activities and do the things they wanted. It was important, not only for people with HD to remain active, engaged and fulfil their wishes, but also for family carers to have enjoyable things to look forward to. For Amy and Martin, their condition had progressed to the point where it would not be possible to go away without considerable carer input. Julia engaged with the Huntington's Disease Association local carers group and Martin participated in some of the trips they had organised. These trips had added support from local families and friends of people affected by HD.

CONCLUSION

As personal ability decreases and needs increase the person with HD slowly moved into a middle phase of their condition. Day care, respite and home care were all introduced throughout this time depending on the person with HD's level of disability. Families were only then able to maintain a balance of caring for someone at home by additional support and input from services. In addition HD slowly encroached on the home space turning it into a hospital at home. Adaptations were made to areas of the house, and equipment and medication were stored. The emphasis of the home environment was shifted to incorporate a constant throughput of people providing different services. Well members of the family may even have become confined to certain areas of the home.

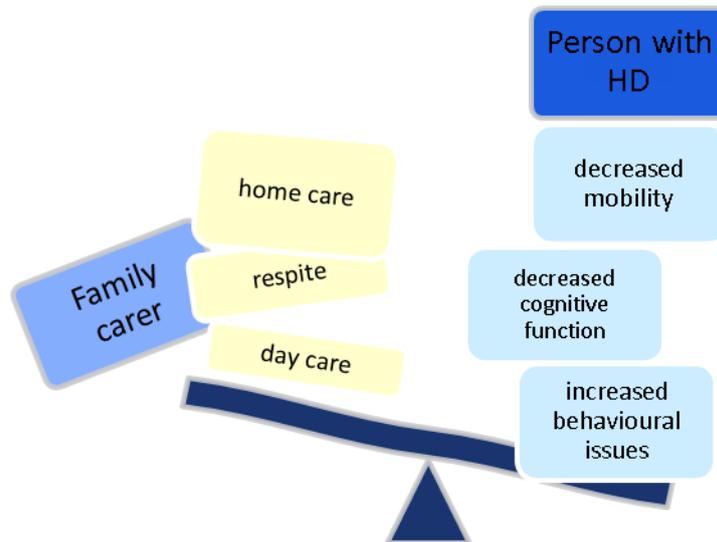
As their condition advanced participants in this chapter talked about the different types of services provide to help them maintain the person with HD at home. These included day care, home care and respite services, not all of which were embraced or utilised by all participants. As far as possible, services were necessarily tailored to meet the personal, environmental and support needs of each of the people with HD.

It was at this point in the disease trajectory that family carers and health and social care professionals continued to balance increased risk of injury or exacerbations and allowing the person with HD to make choices and live as they wish. For example, balancing maintained muscle strength and independence by walking versus the potential injury and loss of confidence from falls; the continued pleasure and normality of oral eating versus the risks of choking and longer term effects of repeated aspirations.

Acting as informal care co-ordinators, family carers took on the bureaucratic processes involved with gaining access to services. The health and social care professionals involved also worked within these systems while trying to provide the best care for their patients. A number of professionals and family carers expressed frustration with these processes, particularly when decisions were made by those with limited understanding of the condition and without consultation. This chapter has also addressed some of the issues raised by

family carers about their decreasing independence and increasing envelopment in the carer role. These changes took on different meanings depending on the familial relationship between the family carer and person with HD. Family carers tried to balance their caring role with other aspects of life such as work, social, and personal time.

CHAPTER EIGHT: RELOCATION AND RECONSTRUCTION



Later phase – balancing relocation and reconstruction

INTRODUCTION

This final findings chapter maps the impact of a move to a residential care home on the person with HD, their family carers, other family members and the health and social care professionals involved in their care. The first section of this chapter examines how a move to residential care is considered to be a last resort. Once out of the family home and in residential care the dynamic of caring is disrupted for family carers who must renegotiate their role in the provision of care. Family carers must also cope with their liminal⁷ status between being a spouse and being on their own at home. The third section of this chapter then goes on to explore the ways in which family carers made efforts to reclaim a life beyond caring. I illustrate how key health and social care professionals remained involved in the person's care after they had moved to a full time residential care home. Health professionals remained key support and advice for the care homes and provided a liaison role between care homes and

⁷ An anthropological concept meaning to be on the threshold or between social states

specialist services such as hospital-based neurology services, physiotherapy or GPs.

Six participants, shown in Table 12, experienced this move to residential care and their cases are used to illustrate the issues raised in this chapter. Four of these participants were living in residential care prior to their start in the study (Kate, Alison, Mary and David). Sarah moved to a local residential care centre after her husband had a stroke during the first few months of the study. Erica's move occurred towards the end of the study period when she had a psychiatric breakdown and was admitted to a specialist mental health unit at the local hospital.

Table 12 - Cases to illustrate issues around the move to residential care

Name	Age at diagnosis	Time since diagnosis at start of study	Family situation	Previous living situation	Time in residential care at the start of the study (date of move)
Alison	54	6	Separated, two adult children	Lived with her daughter until she could no longer cope	3 months (June 2007)
David	37	15	Married, no children	Lived with wife until she could no longer cope	14 months (Feb 2006)
Erica	52	3	Divorced, two adult children	Lived alone until she had a psychiatric break	Was admitted to a psychiatric unit after 34 months in the study, she was still awaiting long-term placement at completion (Jan 2011)
Kate	30	12	Divorced, no children	Lived alone until she recognised she could not cope	4 years (2003)
Mary	40	12	Divorced, three adult children	Lived alone with some home care assistance	2 months (December 2007)
Sarah	55	6	Married, two adult children	Lived with husband until he had a stroke	Moved to residential care home one month after starting in the study (Dec 2007)

THE LAST RESORT

As shown in the previous chapter, as the condition progressed, often over a number of years, increased burdens were placed upon the family carer in the home. Assistance in the form of home help, day care and respite alleviated some of the weight of this caring, however, could not be a substitute for a family carer. When a physical or psychological breakdown of the carer occurred the balance finally tipped and forced the person into alternative living arrangements. In some cases, such as those of Kate and Mary, no family carers were available to assist with any care needs so this shift to residential care occurred earlier than perhaps it would have had a stronger family care structure been in place.

Health professionals recognised that community based supporting services could not provide 24 hour cover at home. The considerable burdens placed on family carers were often at detriment to their own health. Once again the key health professionals participating in this study saw their role as one of pre-empting potential changes in health status and care circumstances. They would introduce the idea of residential care before it was needed and support that decision if it was made by the family. For those living alone it was about introducing the idea of residential care and preparing them for the probability that it may be necessary in the future.

There's also a point at which maybe the people aren't being properly supported at home or it's become more difficult for the carers. I think it's at that point then you have to be very sensitive to everybody's needs but then to think about maybe it's time for going into a nursing home. And quite a few carers have been quite relieved when that's been brought up because they've felt that I've got to stick with this no matter what. So it's giving people those options really. We'll support you to look after your person at home, or there are other options too, and just making those available so they know there is something else if need be. ... I think it's just allowing them the opportunity to say no I've had enough, and feel safe and secure in doing that and not guilty, because guilt's the biggest thing I think with people. As carers they feel guilty. (CNS, Interview 1, 24.06.09)

FAMILY CARER BREAKDOWN

For three of the families in the study it was the physical or psychological breakdown of the family carer that resulted in a move to residential care for the person with HD (David and Jill, Sarah and Max and Alison and her daughter). Cognitive and behavioural issues are often cited as more difficult for family carers to manage than the movement disorder and cumulative physical disabilities associated with the condition. For David's wife Jill, it was David's depression that proved to be the most challenging symptom for her to manage at home.

And then when, a year last October David, the mental health side of it, you got really depressed didn't you? ... And that was the first time through the illness David had never been depressed. And had lots of input from different people, and everybody was very helpful, and CNS got help from the mental health care unit at ... hospital. And eventually after Christmas I was on my last legs because I didn't know how to help anymore, and the [consultant] there said to me, he said 'I think we've come to the end of the road and we need to get David into care'. (Jill, David's wife, Interview1 02.04.2008)

Alison had a short period of living with her daughter after her husband left and it became clear she could not manage her own tasks of daily living. However this placed considerable strain on her daughter to care for Alison, her two young children, maintain a job and run a household. When Alison's daughter could no longer cope Alison moved to the residential care home. Once Alison had settled into the residential care home she was able to reflect and acknowledge that she was not coping well at home on her own and living with her daughter was not a suitable alternative.

I have these terrible accidents and I'm so unsteady on my feet, and I sort of fell, bad, I fell over and broke my nose in three places at half past eleven one night. So that's how I come to be in here ... I was staying round my daughter's. ... Her flat. A bit crowded.... taking [the kids] to school, looking after me ... I was in her bed and she was in her daughter's bed. [The social worker] had to come and see how I was coping. Because the social worker

was in touch with [my daughter] and told her to sort me out because [my daughter] was having a breakdown because she couldn't cope anymore. (Alison, Interview 1, 19.10.2007)

Alison had been offered further assistance in the community. Social Services had suggested she move to a flat opposite her daughter's with an alert alarm in case she fell. However after Alison's recent fall and injury to her nose she recognised her need for 24 hour care and support, despite this not being available in an area close to her family.

Alison: But they said I could have had an alarm, you know an alarm in the flat there, opposite my daughters. Do the flat out for me with an alarm. But sort of having had that bad fall I turned it down (the flat).

Interviewer: Yeah did you think that perhaps you maybe needed a bit more help?

Alison: ...Yeah. Having been there all night long, with a broken nose, I wouldn't have wanted to stay [on my own] ... it's too dangerous. (Alison, Interview 2, 18.02.2008)

In my notes after this interview with Alison I queried whether she should have been offered the living arrangement of a flat on her own. I felt it perhaps reflected a lack of understanding of the condition by the service providers in her area. At her third interview approximately 18 months after moving to the residential care home Alison recognised the importance of the 24 hour care and support she now received.

Well I am quite happy here, I think I made the right decision moving here, because I wouldn't be able to cope with being sick all the time and falling over. See if I fall over here, which I have done twice in the last couple of weeks, I banded my head, but at least they are there to pick me up and that. If I had taken that flat I would have been by myself wouldn't I? (Alison, Interview 3, 18.07.2008)

Family carers often had health issues of their own, particularly those in the older age groups. Several family carers talked of their own health issues

including: heart conditions; arthritis; diabetes; fibromyalgia; and cancer, as well as increasing physical frailty due to advancing age. Hence it was not only the emotional labour of caring that that took its toll but the physical tasks involved, alongside running a household and often working and/or raising children. This was compounded by the persistent nature of the family carer role resulting in interrupted or poor sleep, little time to relax, limited time to themselves, and reduced social activities.

After caring for Sarah at home for many years, in late 2007 Sarah's husband Max had a stroke. Sarah had a care package in place which consisted of morning and evening carers coming in to help with washing and dressing, she also attended day care and had respite time in place. At the time of Max's stroke Sarah was at a residential care home for respite, fortunately the care home was able to arrange for her to remain at the unit.

... the Social Services were marvellous because one call from them at four o'clock on a Friday afternoon, ... but the next morning people were ringing me up on the mobile, leaving me messages and everything to say don't worry, we've been in touch, we've been to [the care home] and they can look after Sarah for the time being. ... And then it became obvious that I couldn't look after her, not only from a physical sense, I mean I'm still a little bit unsteady although I'm far better than I was. And they said look, you've had all the stress and everything for five years, if you ever felt you were able to look after her it's going to happen again. So we made a decision just after Christmas that she would have to stay there. (Max, Sarah's husband, Interview 2, 12.03.2008)

In a later interview Max was able to reflect on his illness and how he felt this incident legitimised Sarah's move to residential care.

I mean it was December when I had a stroke. So that changed a lot ... Oh it certainly made things easy ... So I guess it brought things forward by about, I don't know, two and a half years or so. But it also gave me a handle to hang things on, because I didn't feel guilty about it, couldn't do anything else could I? And I guess we jumped the queue as well, because there are

still no vacancies at [the care home]. (Max, Sarah's husband, Interview 5, 04.11.2010)

UNSUPPORTED

Those participants living alone (Kate, Erica and Mary) predominantly struggled with activities of daily living such as cooking, cleaning, washing, and dressing. Trying to manage these daily tasks became increasingly difficult for some, placing considerable strain on their mental health. For Erica the strain of trying to manage daily living on her own resulted in a severe psychiatric breakdown and admittance to a psychiatric unit. As reported in Chapter Six Erica always contended that she was coping well making it a challenge for health and social care services to accurately judge her care needs.

In January 2011 Erica was brought to the out-patient's clinic for an emergency appointment by her children as both were concerned by her behaviour. I was not aware at the time why Erica was in clinic and when I asked if I could attend the consultation she agreed, as did her children. It became apparent during the consultation that Erica was suffering from some kind of psychiatric breakdown. She was hearing voices and openly spoke to them during the consultation. She struggled to process the questions she was being asked and it quickly became apparent to the consultant and the RCA that she should not go home at this time. I stayed in contact with the RCA regarding Erica's condition. She remained on the psychiatric ward as no satisfactory discharge plan was agreed. The RCA was involved in the discharge meetings and was able to share a better understanding of HD beyond the immediate psychiatric issues.

The discharge meeting re: Erica went ahead as planned. There were loads of concerns about Erica's ability to cope at home as she had fared badly in all the [Occupational therapy/Physiotherapy] assessments. Erica is adamant about going home with help. The community psychiatric nurse is setting up a Rehab team to spend the first month supporting Erica when she returns home and then she'll receive a home care package. I have my reservations about her going home. ... I shared my reservations about the discharge at the meeting which threw a spanner in the works as although

everyone felt Erica has deteriorated no one had considered an alternative to home. She is having weekends at home and then there will be another discharge meeting to discuss her return home/future care needs. (RCA, email correspondence, 04.04.2011)

The RCA felt that the care package being proposed for Erica was not sufficient, making it unsafe for her to return home without 24 hour support. It was not clear if this was due to a lack of understanding of Erica's condition as not just a mental health issue, or the constraints of the community service provision. However, Erica's case demonstrated the importance of family carers and the key role they play in providing a substantial proportion of care to people with long-term conditions in the community. Erica was divorced and her two children in their early 20s only lived at home sporadically. It also illustrates the role the RCA played in advocating for Erica and providing detailed information to those unfamiliar with HD.

Interviewer: So she is really just in [the psychiatric unit] at the moment because there isn't anywhere else for her to go isn't she?

RCA: And everybody keeps offering her ridiculous levels of support you see. ... I think they just need to look for some residential support. ... And you have got to look at that because Erica isn't safe. She would be safe with a live in carer I think, or safer. But no one will fund that these days. They will say no, they are not supposed to look at the cheapest, but that is what happens. They were offering 3 hours support a day for her, well how is that going to work? (RCA, Interview 2, 07.07.2011)

Although not placed as urgently as Erica, Mary had come in to the care home for a temporary respite period. It then became clear that without the 24 hour provision of a family carer she would not be able to return home as a sufficient care package would also not be available to meet her needs. Again Mary was divorced and had children in their late teens and early 20s who did not live at home.

Additional life strains such as family bereavement, divorce and moving home contributed to Kate's 'downward spiral'. With the help of friends she was able to

recognise that she was no longer able to cope on her own and began to look for residential care homes for herself.

Kate: So I realised that I was at the end of my tether, [my friend] said I've never seen you so, you know, I just couldn't seem to think, my health was bad. I felt like everything was getting worse. ... When I came in I just felt relief to be honest ... Really it was a combination of still grieving for my mum and not having had time to get over the loss of my marriage.

Interviewer: So did you feel that you needed the nursing care?

Kate: Yes... I couldn't have made it, couldn't put in to words what it actually meant to me but it's been really good because it's given me a rock solid foundation. And even if I'm not well I know I've got support with the nurses, they're really good. ... seeing [my mum's] specialist helped me know that when it was time to ... accept that my illness had progressed to the point I needed to be here for my own safety then I accepted it. (Kate, Interview 1, 14.06.2007)

Kate and Alison demonstrated good insight into their physical and mental health. Erica displayed a classic symptom of HD by always contending that she was 'fine'. It is not clear how much Erica lacked insight into her deterioration due to HD and how much may have been her choice to deny the changes in her health. She did this during her interviews with me and in the clinic appointments that I observed. This made it hard for health and social care professionals involved in her care to ascertain how she was really coping, leaving them to piece together a broader picture using information from family members. The ongoing difficult relationship with her daughter made Erica's living situation somewhat erratic as her daughter stayed at home and lived elsewhere several times during the study period. In my notes on Erica's emergency clinic appointment I queried whether closer monitoring of her home and behaviour could have prevented her psychiatric break.

... Erica seemed relieved at the end [of the consultation] because when she stood up she had quite a coherent conversation with the consultant so the anxiety seemed to have subsided a bit, maybe because something

is now being done? Makes me think of what CNS said about how she keeps an eye on Helen. Could this have been avoided if someone like the CNS had been in place to visit? I think probably her ability to hide her difficulties meant that in this instance maybe a crisis was necessary to really bring issues to a head and get the care she needs in place. She has always maintained she is fine, despite the consultants' and RCA's best efforts to get her talk more openly and working with her daughter. (notes on observation of Erica's emergency clinic appointment, 25.01.2011)

'NO LONGER A CARER'

For those family members who had not cared for the person with HD at home little change occurred when their loved one moved to residential care, and they maintained their visitor role, usually on a weekly basis. For example Alison's sisters, estranged husband and children all visited her at the care home and often took her out for the day. They would also supply her with toiletries or food when necessary. Mary's daughters would also visit her, as did Kate's friends and Erica's children. However for those who had been family carers and performed the care role at home shifting into a 'visitor' role and fully relinquishing their status as 'carer' was more difficult. Many stayed engaged in a number of ways visiting on a regular basis and often travelling a considerable distance to do so.

Two family carers engaged with the study and participated in a number of interviews and observations over the full three-year period, Sarah's husband Max and David's wife Jill. They talked about what it meant for them when their spouse moved to residential care. Both remained heavily involved in their spouse's care. Beyond visiting and providing essentials they would also help make their spouse's room more homely, attend consultations and often helped with personal care and activities of daily living like washing, dressing, teeth cleaning, hair brushing, shaving, eating and laundry. Both continued to take their spouse out of the residential care home for the day and for the first few

years Jill was able to have David at home for weekends and holidays. Max visited Sarah at least twice a week. Jill also visited David at least twice a week and, as facilities allowed at David's care home, she was able to stay over on occasion. Both also remained involved in care decisions and health assessments. Max almost always attended out-patient clinic visits to the Birchtree team with Sarah and a nurse from the care home. Jill made sure she was visiting when David was assessed by any external health or social care professionals, such as the GP or speech and language therapist. Jill also maintained a close relationship with the care home staff and remained involved in discussions about David's care and well-being. She spent considerable amounts of time at the care home volunteering to help with social and entertainment activities and bringing her two dogs for Pets as Therapy (PAT).

Yet despite this level of input I suggest family carers found themselves in a liminal position regarding their 'care role'. Many of the practical tasks they had performed daily at home had been relinquished to care home staff and family carers queried their new position. After Sarah moved to full time residential care due to his stroke, Max struggled to come to terms with living alone. He felt it was important to remain involved in Sarah's care, to get to know the care home staff, and let them get to know him.

Strange. Learning to live by yourself. There are two sides to it. There's the sort of physical side of it, there's nobody there, there's nobody for hugs and kisses when you need them, and I'm sure she needs them as much as I do, and just having someone to talk to, even if there's somebody there to listen as it were. There was a purpose if you like, I mean I was looking after Sarah, that was my role. ... So I think it's important for me to get to know the people who [at the care home], and they know that I'm Sarah's husband. (Max, Sarah's husband, Interview 2, 12.03.2008)

For Jill handing over caring tasks she had performed for so long was difficult. In giving up their official 'carer' role, family carers struggled to carve out a new position in the dynamic of care provision.

I shall never forget, I came here, I just didn't know what to do with myself really, and I can remember me packing, unpacking in [David's] room and packing again, putting it all back in the case. And you said what was I doing, I said 'we're going home', but you wouldn't let me would you? ... But now David's here, I felt a year ago very upset that he was here, not because it was the wrong place but because I wasn't in control. Although I was really too tired to be in control. But I felt I would be better, giving myself six months or nine months I would feel better, but I'm 12 months down the line now and I'm still the same as I was. ... I'm still extremely tired because I won't let go, and that's my problem. ... handing the reins over to other people is quite difficult. ... I've no qualms that some of the people here do things far better than I could, I've no qualms about saying that, but it's the emotional drain isn't it? (Jill, David's wife, Interview 1, 02.04.2008)

Family carers did not relinquish their role lightly and it took considerable time for them to see themselves as anything other than 'carers'. Three years after Sarah had moved to residential care this liminal status was revealed for Max.

[An] interesting comment that was made when I was at [a meeting] last week was 'because Sarah's now in a full time care home, technically you're not a carer anymore', which is interesting. ... Some of the things they've said we want help here, we want help there and everything, and when I said well if you're short I'll come along and everything, but I can't be involved in everything. But they've said 'well hang on, you're not a full time carer anymore'. (Max, Sarah's husband, Interview 4, 16.07.2010)

Despite having their primary role removed family carers did not relinquish all 'caring' and it could be argued that their burden of care has merely been shifted, rather than actually lifted. Once physical caring tasks were removed this exposed a greater level of emotional burden, one which often found to be harder to manage than the practical tasks of providing 'care'.

But I still want more, trapped in this, ... I feel as if I'm sort of like treading water because you are married but you're not married, I've got a home but half of me is here, and it's just a mess really isn't it? But a good mess if you

know what I mean, he's well cared for and I can walk away and know that when I go home now I can go home and go to sleep. ... I can go home now and know that he's safe, relatively safe, safe as he can be. But the emotional side of it is just awful. (Jill, David's wife, Interview 3, 21.04.2010)

Initially Jill was able to continue to perform some physical tasks but as David's disabilities progressed she had to relinquish more and more of this physical involvement in his care. When David had been able to come home for weekends and holidays during the early parts of the study period Jill had still been providing full care for him during this time. As it got to the point when she could no longer physically manage to lift and move David these home visits ceased. At this stage it seemed as if her role shifted from the active provision of 'care' to a more passive 'caring'.

It's tiring, and it's not because I'm, most of the time I'm not physically doing a great deal now whereas I was before, but now I can't do a lot because I can't even move him, whereas before I would get him ready to go out and move him and get him in the wheelchair and take him out. I can't do that now, but I find it more tiring actually. Sort of sitting and chatting a bit and reading a book or doing some sewing or something, well I find it quite tiring. ... because I can't do anything. If you can do something positive and you can see that that's made a difference [and] I'd feel better somehow. (Jill, David's wife, Interview 4, 01.06.2011)

Max had tried to maintain many of the things Sarah liked to do outside of the residential care home by taking her to get her hair done, to the park, to restaurants, and the cinema. Similar to Jill's experiences Max found this care role diminished as Sarah's physical disabilities increased and she required additional help to be moved. This meant that Max could no longer take Sarah out on his own, eroding another practical element of the carer and family member role.

But I mean one of the big things is that she can't walk anymore, and she can't even stand, she can't hold her own weight, which means she has to be hoisted everywhere, which means that we can't transfer her into the car. So

me taking her out in the car has been, probably for about four or five months now, not an option. And I'm waiting to become a van driver because I've ordered one of these cars which is wheelchair access through the back, so that at least I can take her out by myself, whereas at the moment it's with the goodwill of [the care home] and I can't go without a carer, I mean I just can't cope on my own anymore. (Max, Sarah's husband, Interview 5, 04.11.2010)

As Sarah's condition deteriorated over time Max had to make a shift in his understanding of his role. As the study period continued it became apparent that Max was slowly being able to relinquish more of his carer role. Max was on holiday when a clinic appointment had been made for Sarah, this seemed to be the first time he was happy for the care home staff to take Sarah to the appointment without him.

And I said there's nothing I can do when I'm there (at the clinic), because really [the care home staff] see her on a day-to-day basis, I see her two or three times a week, so I can add my four penn'th but they see her on a, for every three hours that I see her, they see her for three days as it were. (Max, Sarah's husband, Interview 4, 16.07.2010)

Understanding that this shift needed to occur was one aspect of the transition the family carers were undertaking. To embrace a new or recharged role as an individual away from the care home was a further challenge and required a shift in attitude toward 'not giving up on life'.

'NOT GIVING UP ON LIFE'

It was important for the person with HD not to give up on living as full a life as possible. Family members spent time taking them out and care homes provided a range of activities for all residents. For example Kate was part of a riding for the disabled scheme and was able to go out once a week to ride horses. Activities provided by the residential care homes also included arts and crafts,

music, computer games, therapy games, entertainment shows, tea parties and Pets as Therapy. Staff at the residential care homes recognised the importance of the activities and entertainment programmes and were often frustrated that they lacked sufficient staff to provide this element of care as comprehensively as they would like, particularly on a one-to-one basis.

The fact that we cannot physically in this environment, in this time, we're always short staffed; we've never got enough staff. So you haven't got enough staff to do the job and do the fun. In an ideal world we'd be slightly over staffed so that you'd have one person that could read a paper, paint a nail, do a makeup, whatever, and make time to have that kind of interaction which we don't have. (Staff nurse Larch Tree Care Homes, Interview 2, 29.07.2010)

Family carers too tried to remain social active throughout the time their spouses had been in residential care. Max was a regular walker and bird watcher and participated in a number of carers groups and supportive organisations. Jill initially continued to work when David moved to the care home, but also had an elderly mother living with her and participated in volunteering at the care home, along with an advisory role in the UK Huntington's Disease Association. Jill had also trained two spaniels to become Pets as Therapy dogs and regularly brought them to the care home for David and the other residents to enjoy. These activities were their way of contributing to the HD or health care community and undertaking activities which drew on their carer role in a different way. It was the idea of personal time and a personal life as an individual that challenged them to re-assess their identity as a carer. Both Max and Jill latterly gave up their roles within the HD organisations, partly as Sarah and David deteriorated, these became additional burdens, but I suggest this also served to disengage them from some aspects of the HD community as they tried to move on with their lives.

Family carers slowly embraced the realisation that they had to move on or 'away' slightly to regain some of their own life, but that this would be with a shifted identity. They were no longer part of a couple but still had a spouse.

They had to reintroduce themselves to a social world and potentially one of work and sustaining themselves. However this was while maintaining a relationship with the person in care and continuing to tend to their needs.

I think it's been my psychological process through this that's been [the hardest], for me it was always David's psychological thing that was the biggest problem for me, because the physical I could do something about but psychological you've got no control over have you? So that was really hard, but me being in the position where you are, you've got somebody but you haven't got somebody and it's just that really weird sort of feeling. (Jill, David's wife, Interview4 01.06.2011)

Remaining busy and active was often a distraction from the negative and undeniable future of their spouse. In his final interview Max talked about some of the changes in Sarah's condition over the previous few months. Staff had begun to discuss the option of artificial feeding and Sarah was now unable to weight bear. Max recognised that as Sarah's condition deteriorated she was beginning to near the end of her life.

So there's been a big big change from that point of view. And I suppose from my point of view, the realisation just to get on with your life, it's not bad. I mean I've been away. ... I've been to Scotland trekking in the Cairngorms for a week and things like that. So that's helped and kept my mind off things, but I must admit things are starting to concentrate the mind. (Max, Sarah's husband, Interview5, 04.11.2010)

Jill was not as far along in her transition as Max, and in her final interview she reflected on the time David had spent at the care home. Jill talked about how David's time in the care home had impacted on their relationship and her attitude towards her continued liminal position between wife, carer and individual. David had recently been quite ill and suffered a serious fall at the centre whilst Jill had been on a short holiday. I observed the changes to his room in my fieldnotes on my final visit.

Same nice, homely room as before but stripped down to basics to stop David from hurting himself. Bed in lowest position with crash mat on the floor next to it, padding on the wall and along the edge of the bed. Only other furniture now is his specialist easy chair and the wardrobe, and TV on the wall. Desk, chair and all other furniture removed. (notes on care home visit with David and Jill, 01.06.2011)

Despite his fall David persisted in trying to get up unaided and fell many times. This had resulted in staff organising 15 minute checks and stationing a staff member outside his room. Jill had also spent considerably more time at the centre. This incident and intense period of concern clarified for Jill that despite every one's best efforts to keep David safe and well things could change at any time. This knowledge seemed to allow her to try to reduce the intensity of her links with the care home and try to re-establish a life outside that of being a 'carer'.

You go along at a level for a certain amount of time, and it was difficult coming [to the care home] and being used to David not being at home, but I had the opportunity to bring him home sort of every other weekend and things, ... So we came here and had Christmas here. But all those big major events are milestones aren't they? And now I look back ... David was eating, ... now he's not eating at all, it's just changed so quickly. ... So my psychological things change as well, and I don't know how, I think just trying to let things evolve ... But I'm doing more things in the community now, I've joined more things. What I found difficult I suppose being married for so long is not being a couple, not going to the things that all the people that we've been with so long are going to, you just don't, it's just life changes. And just this last two or three weeks, ... because I don't know what I want to do really. But I am doing things. (Jill, David's wife, Interview 4, 01.06.2011)

Max has two adult sons and by the end of the study period had three young grandchildren. Despite none of them living in the area Max remained engaged with this wider family.

One of the ladies lost her husband here just a couple of weeks ago, and she has children, adult children, and so her life is very different. ... she's involved in other things that are going on with them. And I thought yeah it's very different when you've got your unit. I haven't got that, so I've got to get out there and do things, it's not going to come to me, I've got to go and do it. ...I don't want to give up on life, that's how I feel, I don't want to give up on life. David's world is so small now, and I protect him to the nth degree but you can't go down that road can you, it's not my life. But it's very hard to step away from that and not feel guilty. (Jill, David's wife, Interview 4, 01.06.2011)

As David and Jill researched David's father's HD early in their marriage they agreed not to have children. For Jill now this means she does not have her own family 'unit' to help her re-engage with a social world outside of one based around HD.

THE CONTINUING ROLE OF THE HEALTH PROFESSIONAL

For those who had links with key health or social care professionals in the community, these remained in place once the person had moved to residential care (Sarah, David, Erica). All the care homes involved in this study had considerable experience of people with HD. However they still consulted community based professionals for their advice and clinical opinion. Each person with HD had different symptoms and affectations. Professionals in the community often knew the person for a number of years as well as having access to a wider range of health and social care professionals and were often used to liaise between care homes and consultants. They also helped discuss difficult topics with families, acted as an advocate for the person with HD and/or their family carer and were utilised as a source of knowledge and expertise. This seemed to be an important relationship both for care homes and the community based staff. Community staff would provide education sessions for care home staff, and care homes that were perceived to be able to cope with the complex needs of people with HD were well known to community staff. This

is a brief extract from my observation of a meeting between Jill, David's wife, the staff nurse from his care home and the CNS for the HD service in David's local area.

Also discuss hoist (CNS is translating Jill's concerns to the staff nurse, she does this with authority but with respect for their roles and in a polite way). (notes on observation, care home visit with David and Jill, 01.06.2011)

The Larch Tree care centres recognised they were self sufficient, often due to their physically isolated locations. These care centres were situated in different geographic locations across England where community based resources for HD varied considerably. The fact that the CNS from David's local service travelled beyond her assigned area to continue to visit him at the Larch Tree care centre seemed to be a fairly unique situation. Larch Tree care homes did not report other visits from HD specialist nurses and this may be due to limited number of these roles in the community. The CNS for the Birchtree team maintained strong links with the local care homes in her area and continued to see all patients once they moved to a local care home. However, as the CNS was one of the key participants in this study I was able to gain a greater understanding of her role. It is not clear whether others in CNS roles for long-term conditions would also maintain links in this way.

It was evident for Sarah and David that their local CNSs remained involved in their care and often acted as advisors to care home staff. Sarah still had regular appointments at the out-patient clinic in Birchtree, as she had prior to moving to the care home. The speech and language therapist and the dietician would also visit her at the care home along with a number of other residents and respite clients. This meant that Sarah still had the same access to the CNS, speech and language therapist, dietician, consultant and Social Services as she had done at home. Mary also reported that the RCA from her local area kept in touch and visited her when she initially moved to the care home. It was not clear how long this contact was sustained for. The RCA in Oakfield remained a key component of Erica's care after she was admitted to the psychiatric unit, mainly

taking on a role of advocate and expert advisor for those trying to provide the most appropriate care for Erica.

For those in Larch Tree care centres their health was monitored by the local GP in the first instance. Referrals to speech and language therapy, physiotherapy, dietetics, and mental health services were made as and when required. Kate and Alison did not report any links with specialist HD professionals in the community such as a CNS, prior to their move to residential care. Links with a hospital out-patient clinic Alison attended while living at home were maintained and she was reviewed by their neurological team approximately every six months, either at the hospital or the care home.

One of the staff nurses from the residential care home where Sarah lived recalled efforts to resolve the issue of Sarah's refusal to eat. Sustained links with the community HD team were considered essential for advice, support and clinical review when residents with HD needed additional input at the care home. A key element of this working relationship both between care home staff and community health professionals and with residents was the continuity of this care. The community team were well known to the care home staff and the residents with HD.

It takes a lot of people to feed her sometimes. Sometimes we need three people swapping over in a meal time. I did everything, I had speech and language in, I had dieticians in. I had the GP in to check if there was any underlying cause, is it her teeth, is it her mouth, is it this, is it that ... And I do work really closely with [the CNS] and [the dietician] and [the speech and language therapist]. ... I do prefer to go into [the out-patient] clinic with [the residents]. I like to follow them through the lot. (Staff nurse, a Birchtree care home, Interview 1 24.07.2009)

It was clear that the community HD team were very much still involved in Sarah's care after she moved to the residential care home. Since Sarah's move to the care home Max reported having little contact with the CNS outside of Sarah's clinic appointments. However, having built a relationship with the CNS and the community based team over a number of years Max felt confident in contacting

them if he needed help and it seemed to be the knowledge of their existence that he found reassuring and supportive rather than any particular need for action at this time.

But I mean I don't have a great deal to do with [the CNS] now, I see her at clinic and apart from that, or we need her help on special occasions like when the continuing care assessment comes, it's absolutely essential to have [the CNS] there because the people who do it don't understand. ... it's a great comfort to know that there's a team around you. You don't have to use them every day and everything, but you know they're there and if something happens you know where help is. (Max, Sarah's husband, Interview 5, 04.11.2010)

Despite David's care home being in a rural area and some distance from his home in the community, the local CNS maintained contact with David, his wife and the care home. The CNS was able to liaise between the care home and specialist hospital-based staff as David was no longer able to travel to out-patient clinic appointments. For Jill this was not only a key part of her support system but an important part of David's care, particularly as this relationship had been in place for almost a decade.

She keeps coming because she's the one that sort of liaises between specialists. Now David's not going to hospitals and things, she will liaise with the specialists for medication and she'll make recommendations ... It's really good with that contact with [the CNS] isn't it David?, because we've got that contact with the past and the continuity is good. (Jill, David's wife, Interview 3, 21.04.2010)

The longevity of David and Jill's relationship with the CNS became increasingly important as his condition deteriorated. David's recent visit to hospital due to his fall prompted him to update his previous Advance Directive stating his wishes, specifically that he did not want any hospital admissions due to his condition. Jill requested a meeting to discuss these issues with David and the health professionals involved. The GP queried David's capacity to make such

decisions and the CNS was then consulted to provide contextual information and knowledge of David's earlier decisions.

[the CNS]'s been in as our link, and she's been assessing it all the time since David was early diagnosed at home, and she's just there all the time. They go from [the care home] to her and we go to her. It's very good to have somebody that's got that background knowledge that knows what's happened. ... David had made this advanced directive quite some time ago, and I didn't know how that would apply now, because things change so much don't they. ... [the CNS] was the one that we did it with at home and she was our representative. So again that link has been really good to still have there. ... because she could think back and look back in her notes of what David wanted at that time. (Jill, David's wife, Interview 4, 01.06.2011)

It was evident from observation of the CNS's visit that the care home staff were responsive to her advice and that Jill was reassured by her support. On her visit to the care home she spoke with David and assessed his comfort and condition, she then liaised with the staff nurse on duty and spent some time talking with Jill in private. My fieldnotes reflect the essence of that conversation. Jill seemed to want to establish if she had the correct understanding of how much David's condition had deteriorated.

Jill wanted to know if they are 'on the same page about how things are at the moment' (this seems to be in relation to David's health and general care). The CNS agrees they are and acknowledges that there have been a number of changes in the last 6 months. ... The CNS says 'It is coming to that stage where he may get an infection and may not be able to fight it off' but she agrees with David and his wife that it is right for him to stay at the care centre as he 'can't handle different situations'. ...The CNS says 'it matters about his comfort and safety, keep his quality of life going, that is different from what it was, so just laying with the dogs is enough'. (notes on observation of conversation between David's wife Jill and his local CNS, 01.06.2011)

I noted that Jill seemed reassured by the conversation. It seemed to help Jill to talk through these issues of David's end of life care with someone outside the care home staff. I would suggest that continuity of this relationship built trust so that Jill placed considerable value on the opinion of the CNS.

CONCLUSION

This chapter has examined those in the study who had moved to 24 hour care units. From the discussions with those with HD and their family carers it became evident that moving to residential care was often a last resort when balance could no longer be maintained in the community by supporting services and family carers. Community based services cannot provide 24 hour care, hence family carers are essential in allowing someone with HD to remain in the community and in their own homes. Some form of breakdown in the family carer system immediately prompted a move to care.

It was also clear from interviews and observations with family carers that once the person they cared for moved to a residential care home they entered a liminal state. They were caught between their status of providing active 'care' and moving to a more passive 'caring' role, shifting their burden rather than lifting it. This shift occurred slowly and was still in transition for these family members/carers after the completion of the study. Family members/carers also struggled to adapt to a new social role of being on their own yet still being so attached to their spouse. This liminal position challenged their abilities to engage with an individual social life and an independent life away from the care home.

We also see for those patients who had well established links to community staff these links were maintained after their move to residential care. These health care professionals also continued to support families in finding a new balance in their role as carer. Specialist professionals with a better understanding of HD, such as those in a CNS or RCA role, were also called upon by the care centres for information and advice. These specialists remained as a link to community and

hospital services such as out-patient clinics and neurological consultants. These ongoing relationships built a continuity uniquely important for the quality of care of the person with HD and their family members.

CHAPTER NINE: DISCUSSION

INTRODUCTION

In the previous chapters I have taken a temporal approach to the presentation of data, using cases that represent themes from the early, mid and later stages of the condition. These stages were not fixed and some participants deteriorated during the three-year study period. This chapter draws together the evidence presented in the three qualitative findings chapters. It is divided into two main sections. The first part focuses on the issues for people with HD and their family carers, and the second on the issues for health and social care professionals. The concept of balancing a delicate equilibrium to maintain the health of the person with HD with that of the family carer is a thread that runs throughout this discussion.

The first part of this chapter discusses how people with HD and families responded to the new knowledge of the diagnosis and the burdens this placed on people to seek and share this information. I then discuss the transformation of home spaces into care spaces as equipment, adaptations and professionals enter this private sphere. Family carers are identified as key agents of care provision and essential to people with HD remaining at home if they wish. Even when a person moved to residential care the burden for family carers is merely shifted rather than lifted. Finally I discuss the issue of choice and decision making suggesting that the professional's role in directed decision making is underestimated and should be acknowledged and embraced. This is presented together with a concept of relational autonomy to discuss the challenges faced by professionals and family members when caring for someone with diminishing and altered capacity.

The second section of this chapter focuses on the health and social care provision for people affected by HD. In light of the findings of this study and in the context of the literature and current policy, I summarise care provision for HD by presenting a model of care. This views the patient, family carer and key worker as a triad of care. The model shows that as the condition progresses

over time the need for care increases. Suggestions are made for the types of multidisciplinary input that might be needed throughout the course of the illness. In this second part of the chapter I explore how quality care was delivered at the three sites in this study. Emphasis is placed on the key worker role for people with HD, how to establish continuity of care and built specialist knowledge. Whether self-management is a valid concept for people with HD is also considered.

PATIENTS AND CARERS

RESPONSES TO NEW KNOWLEDGE

Participants in this study had both emotional and functional responses to their diagnosis. All were shocked by confirmation of their diagnosis despite some having knowledge of HD in their family (Codori and Brandt 1994), and all had to rebalance their lives in order to adapt to this new information or confirmation. Functional responses included making adjustments to their lives from simple things like sitting down to get dressed, to stopping work or giving up driving. Despite being considered in the early stages of the disease all the participants represented in Chapter Six 'Knowledge Management' had presented with motor symptoms which may suggest they had other cognitive symptoms of the condition for many years prior to their diagnosis (Harrington et al. 2012). Such cognitive changes may be subtle but would affect multi-tasking, speed of thinking and co-ordination, things essential for driving and maintaining a number of jobs.

Writers on chronic illness recognise it as a disruption incorporating a number of significant losses (Bury 1982, Williams 2000, Locock et al. 2009). Despite such significant losses to identity and independence, the majority of participants adapted to these changes without sustained disruption. Charmaz (1995) suggests such 'repair' occurs when the person adapts and alters their life to accommodate such losses. Due to longevity and permanency of a diagnosis of HD such intrinsically negative concepts as disruption indicate a 'destructive

breach in the fabric of life' (Locock et al. 2009, p1045). Whilst biographical disruption may be a valid concept around the period of diagnosis and coming to terms with the illness for those with HD, Williams' notion of 'normal illness' (2000, also see Robinson 1993) may be more appropriate in the longer term as people are slowly able to rebalance their lives.

Participants in this study illustrated many instances of what Brown and Addington-Hall identified as a 'sustaining narrative' (2008). In the sustaining narrative, people talked of keeping active and engaged, and keeping what was positive in life. Participants in this study echoed these sentiments by talking about engaging with friends and family, organising trips to do the things they enjoyed and trying to maintain health by keeping active. Overall, participants reported maintaining the normal tasks for daily living such as continuing to undertake decorating, and gardening, whilst incorporating clinic appointments and home visits from health and social care professionals. That is not to say that participants did not have periods of coping less well or struggling to manage the implications of their condition. It should also be recognised that those overwhelmed by their illness and unable to adapt in the longer term may not have wanted, or been able, to participate in the study.

For family carers adapting to the new knowledge of a diagnosis of HD was challenging. All had to adapt their own behaviours in order to accommodate those with HD. Education played an important role in their transitions to acceptance as it was only once they understood the role of the disease in their loved one's behaviour that they were better able to manage and accept the disruptions it caused. This education was provided for some in an information and support day organised by the Birchtree HD team and more generally through their interactions with the HD care providers and home visits from the CNS and RCA. A recent study conducted in the Netherlands demonstrated the positive effects of a patient education programme for those affected by HD (Campo et al. 2012). The formal education programme was provided for both patients and family carers. Findings showed improvements in behavioural symptoms and anxiety for those with HD and less psychosocial burden for

family carers (Campo et al. 2012). This is explored further in the section on self-management later in this discussion.

BURDEN OF KNOWLEDGE/OBLIGATION TO BE INFORMED

Several participants in this study talked about attempting to control information by deciding if, how and when they told people about their diagnosis, the repercussions this has for them, their family, and their social circle. Also recounted were stories of how secrecy and silence within families, both historical and current, impacted on experiences of finding out about, understanding and living with HD (Forrest et al. 2003).

There is limited research on the psychological impact of living with a genetic disorder such as HD. However, some evidence suggests secrecy, silence and stigma still surround this relatively rare and often misunderstood neurodegenerative condition (Hans and Gilmore 1968, Paulsen 2008, Halpin 2011, Almqvist et al. 2001). Several papers have explored the impacts of genetic testing (Meiser and Dunn 2000, Williams et al. 2000, Codori and Brandt 1994, Codori et al. 1997) and a few have investigated adapting to being diagnosed as being at-risk of the disease (Pakenham et al. 2004, Hagberg et al. 2011). These studies focus on why people chose to be tested or not, the stress of undertaking genetic testing, and living with the outcome. Little emphasis is placed on what people subsequently do with this new knowledge or their longer-term coping styles. Some families may use secrets as a way of coping with possible genetic inheritance issues and as a denial or avoidance strategy (Paulsen 2008).

However there are issues of ownership of information and the person's right to choose who to tell about their condition as well as how and when to do so (Lucassen and Clarke 2007, Forrest et al. 2003, Hakimian 2000, Forrest Keenan et al. 2005). Forrest Keenan et al. (2005) also raise the issues of other family members' entitlement to genetic information, including non-biological relatives who may consider themselves to have a 'legitimate interest' (p210) in such information. They highlight that

biological relatives may be denied the opportunity to seek medical input if they are not aware of their genetic risk (Forrest Keenan et al. 2005)

Secrets were often viewed as controversial, with someone taking the decision to hide information from others. This implies a deliberate act and for these purposes is distinguished from silence. Secrecy was maintained for a number of reasons. Participants identified guilt, blame, denial, and coping as just some of the elements involved. In Paulsen's guidance provided through the Huntington Society of Canada she suggests 'Denial or secrecy may be chosen because they are not ready to accept the diagnosis or the fear of discrimination in such areas as employment, promotion, adoption, insurance or social acceptance' (2008, p30). Yet it is the hereditary and dominant nature of HD that, coupled with the dynamics of family and culture, makes disclosure particularly complex.

Forrest Keenan et al. (2009) explored how thirty-three young people between the ages of 9-28 found out about their family history of HD. Their study identified four ways of finding out; having always been told, being told gradually, it being kept a secret and HD being a new diagnosis. Five of the thirty-three participants reported HD being kept a secret from them and recounted the impact of learning this information. A further five recounted HD as a new diagnosis, despite a knowledge of some illness in a previous generation. This suggests either misdiagnoses or the information that it was HD was not discussed and shared with the family (Forrest Keenan et al. 2009). Blocking information in these ways was identified as being horizontal, between generations, and vertical, across generations to protect others. Like some of those in Forrest et al.'s (2003) study, participants in this study often felt that they did not have the authority or that it was not their place to tell other members of the family. However, this was not universal as some felt that arming people with information was more important.

This study identified that seven of the 15 (46%) participants had no knowledge of HD in their family prior to their diagnosis. This reflects Almqvist et al.'s (2001) study in which 44% of participants had no family history of HD when they presented for genetic testing. In my study five subsequently traced their

genetic inheritance and attributed a lack of knowledge to the early death of one or both parents. One cited prior misdiagnosis within the family although this was also identified by several participants as a source of delay in diagnosis and information transfer in previous generations (Halpin 2011). One was not able to identify the line of inheritance suggesting non-penetrance in her affected parent, non-paternity, or a new mutation.

Despite welcoming knowledge and information about some aspects of HD, a number of participants in my study disengaged with the type of proactive knowledge sourced by genetic testing. Conversely other participants made a number of decisions based on this knowledge, particularly reproductive decisions. Huniche (2011) draws on an empirical study of the everyday lives of people with HD and suggests that the availability of genetic knowledge and the resources to explore this knowledge now exist. It is services such as predictive testing, prenatal testing and pre-implantation testing that create these moral issues. Yet participants within my study did not actively engage with the majority of these services and several chose to ignore the knowledge of their genetic risk and continue to expand their families. Just as highlighted in Huniche's paper, some participants in my study chose to 'live outside the public arena of hereditary disease' (2011, p1812), by choosing to deal with 'concrete manifestations' of disease as and when they happened. This recent work by Huniche (2011) and also Weiner (2011) raises wider issues of responsibility and moral obligations embedded in the sharing and transference of knowledge about genetic inheritance. Weiner's study of hereditary raised cholesterol suggests this responsibility is twofold; first there is an obligation to be informed of one's own genetic risk, and to take precautions to monitor and modulate that risk; second to extend that responsibility to other people, especially family (Weiner 2011). It could be argued that in a country with a public health care system a social obligation also exists to minimise the reach of the condition due to the potential drain on public resources which is likely to occur once symptoms manifest.

Fear of stigma and discrimination was identified by participants as one of the underlying reasons for non-disclosure of HD within a family and beyond.

Several participants in this study identified the secrecy and silence being maintained within their families as affecting their knowledge of HD. They also talked of their choices not to disclose their illness to others in order to have some control during a time when many must have felt they had little. Some specifically talked about the changes that had occurred in perceptions, treatment and services over the decades since their relatives' illness, making them re-evaluate their expectations of health professionals. Tibben (2009) suggests that family narratives about members with HD and the input of health professionals are built and passed on over generations and can contribute to negative expectations and stereotypes of care.

HOME HOSPITAL

As the disease progressed those with HD required increasing levels of care input and growing amounts of equipment and supplies. Again these were elements illustrated by the stories of participants in this study and in my own observations over the three-year study period. In these ways the meaning of 'home' is altered and what were once family spaces become sick spaces. In this context residential care homes were also considered to be the person's 'home', particularly their own room. This reconstruction of the 'home' as a care giving space is often discussed in the sociological literature in relation to the geographical body (Dyck 1995, Wiles 2003, Williams 2002, Dyck et al. 2005). Dyck et al.'s (2005) Canadian study focuses on those receiving care and the impact of this care on the home. Dyck et al. (2005) suggest that having care services entering the home challenges its private-ness and the identity of those living there. As the person's abilities decrease their geographic spaces are increasingly confined (Dyck 1995) and the home becomes the nucleus of everyday life. It is with such constrained mobility and independence that increasing levels of services are required and the home becomes a progressively more public, rather than private, space (Dyck et al. 2005, Wiles 2003).

It was not just in a metaphorical sense that homes were reconstructed to accommodate paid caregivers but also literally. Adaptations were made to the home to accommodate decreasing abilities of the person with HD, such as the

installation of wet rooms and downstairs bedrooms. Homes also stored supplies and housed equipment (Wiles 2003). In one instance in this study a room was effectively stripped of furniture and ornaments to reduce injury in the case of a fall (Wiles 2003). These changes in what was experienced and perceived to be the concept of 'home' can lead to a change in the relationship between the person with HD and the family carer (Williams 2002).

These physical changes were clearly observable as this research involved visiting people at home. However, participants rarely referred to a loss of homeliness or an invasion of privacy. Coping with sometimes quite dramatic changes to their home to accommodate the person with HD appeared to be part of the normalisation of the illness (Robinson 1993) with participants seeming to view these changes as practical necessities to facilitate their loved one remaining at home. These adaptations and changes to the home were part of what allowed the input of care to remain in balance with the need for care. A progressive long-term condition such as HD poses a number of questions about the impact of altered home spaces over extended periods of time including the impact on family life, and other people living in the home, such as children. For those in residential care homes adaptations to spaces took place as far as possible to accommodate individual needs. However, the residential care homes were often constrained by availability and appropriateness of spaces and needed to consider the safety and well-being of other residents.

FAMILY CARER BURDEN SHIFTED

As highlighted throughout this discussion and the thesis as a whole, family carers played an essential role in the care of people with HD. Family carers were the predominant way to provide constant and consistent support in the home environment. Round the clock care cannot be provided by the health service and private care of this sort for the extended periods needed for a person with HD is rarely affordable. Earlier in this discussion a concept of normal illness (Williams 2000) or normalisation (Robinson 1993) was employed to demonstrate how participants adapted to the initial disruption of diagnosis and living with HD. However as the disease progressed normalisation was no longer possible. This

study has identified that a lack of family carers, or the breakdown of the family carer, is the point at which the balance of care is tipped and the person moves to a residential care home. I suggest that at this point the challenges and complexities of the condition have taken over and life is identified as 'problem saturated' (White and Epston 1990). It is by the person with HD moving to a residential care home that carers are then forced to start to recreate some kind of re-balance in their lives.

Those participants living in residential care, or who moved to care centres during the study, posed several challenges for formal long-term care providers. Each needed tailored care in terms of; the management of behavioural disturbances, diet, and adjustment to increasing disability. These are similar to the types of characteristics identified by Nance and Sanders (1996) in their study of individuals with HD in long-term care. A further study by Wheelock et al. (2003) identified more severe motor symptoms as a predictor of nursing home placement for people with HD. Those living in residential care homes also had greater psychiatric and behavioural impairments than their counterparts living at home (Wheelock et al. 2003).

Patients in this study living in residential care and their families reported on the high quality of care received from nursing staff and support workers. Family carers remained involved in a range of ways after the person moved to residential care (Skirton and Glendinning 1997). Many would supply food, clothes and toiletries as well as visiting regularly, taking the person out and at times volunteering at the care home (Martin et al. 2008). For those previously heavily engaged in the role of family 'carer' a move to institutional care signalled a transition to a liminal state. This study has provided an insight into the shift in carer burden when the person they care for moves to a residential care centre. Family carers' identities were immersed in undertaking the physical tasks of being a carer. Despite experiencing some relief participants reported finding it challenging to relinquish their previously full-time care roles. As much of the physical tasks of caring were taken over by staff, spouses struggled to shift from the more active role of providing 'care' into one of a passive, but potentially emotionally demanding, 'caring'.

There is some evidence exploring the difficulties faced by relatives once a loved one moves to a care home. Much of the focus is on the care of older people (Davies and Nolan 2003, 2004, Sandberg et al. 2001). However, there is a small body of literature that considers those fulfilling a full time carer role prior to the move to care (Bond et al. 2003, Munck et al. 2008, Kellett 1999, Martin et al. 2008). The move of a partner to residential care left family carers in this study in a liminal state in several ways. Alongside trying to disengage with their care role (Munck et al. 2008) spouses also had to negotiate how to relocate themselves in society as an independent person despite still being part of couple. They were caught between rebuilding their lives on their own and still maintaining a relationship with someone who lived elsewhere and would not be returning home or to a state of health (Martin et al. 2008). This left them balancing grief with the need to move on. Spouses were grieving for an old life with someone who had changed considerably but still wanted and needed to engage with that person. They also needed to move forward and rebuild a life of their own (Bond et al. 2003, Fulton et al. 1996, Sweeting and Gilhooly 1990). Family carers therefore seemed to be dealing with being in transition between their old social position as a 'carer' and the shift into a new social state. In their study of young people with cystic fibrosis moving from child to adult services Tierney et al. (2012) drew on the concept of liminality and refer to 'acclimatising' as the person tries to deal with the transition from their previous identity. Tierney et al. (2012) suggest that a way to reduce feelings of uncertainty during the transition might be to visit the service prior to the move to adult care provision. For those caring for someone with HD, this caring period often lasts for many years making the transition to a residential care home considerably challenging. Time spent at the care homes on respite and day visits could be explored as a way of reducing the enormity of the transition to institutional care for both the patient and family carers. However, community service providers may need to consider ways of continuing to support family carers once a patient has moved to institutional care.

Despite the challenge of dealing with this uncertainty and liminality Bond et al. (2003) found an improvement in mental health once family care givers

relinquished care. Widows of people with dementia living in residential care also had improved psychosocial outcomes and the authors suggest this may be due to a form of anticipatory grieving (Bond et al. 2003). Those that yielded their caring role to formal caregivers following a partner's move to residential care also showed an increase in social activity and a decrease in domestic chores. However, in line with the findings in this study Bond et al. (2003) conclude that despite fewer demands on their time, previously full time family carers 'do not truly exit the care giving role' (p2393) once their spouse has moved to residential care (Martin et al. 2008, Kellett 1999). Hence it is important for families to remain included in care decisions and be encouraged to engage with aspects of the physical caring role as described by participants in this study.

AUTONOMY, CAPACITY AND CHOICE

The Mental Capacity Act (2007) states that capacity should be assumed unless proven otherwise. Guidance is also provided on what may be considered 'unwise' decisions and for decisions to be made in a person's best interests (Department of Constitutional Affairs 2007). However, I suggest conditions such as HD pose a particular challenge for capacity and decision making as understanding and cognitive function diminish slowly and can be coupled with behavioural changes. In the light of the effects of prodromal changes in the brain on the cognitive and psychiatric symptoms (Harrington et al. 2012) and the ongoing cognitive decline displayed throughout the course of the condition closer attention needs to be paid to issues of autonomy, capacity and decision making. This study has illustrated that knowing how and when a person with HD was considered unable to make their own decisions was a process. Therefore decisions were often approached in discussion with all relevant participants in the patient's care. The particular challenge for health and social care professionals becomes whose autonomy is, and should be, given precedence if patient's decisions conflict, or are to the detriment, of the family carer. This study has also highlighted that care decisions are often taken in conjunction with health and social care professionals and are necessarily constrained by the resources available.

I suggest that evidence from this study shows that health and social care professions engage patients and family carers in what I have termed 'directed decision making'. Patients and families were encouraged to make realistic decisions based on available resources and valid options. It was, therefore, a 'steered' choice. Decision making and service user choice are hailed as the gold standard of care provision in service driven NHS. Little attention is paid to the validity or reality of those decisions or choices (Barnett et al. 2008). Findings from this study suggest that directed decision making is necessary and appropriate to guide patients and family carers towards realistic options, and which ultimately can be implemented. Within a public health service 'choice' is an ideology that belies the reality of limited resources. All choices must be bounded in some way and it is the experience and knowledge of health and social care professionals that can enable patients and family carers to be directed towards appropriate and feasible options. I suggest this kind of direct decision making also removes some of the burden of decision making (Ho 2008), particularly from family carers, when the person with HD is no longer able to make decisions for themselves.

We often think about capacity in terms of care planning and end of life care, and Simpson's (2007) paper addresses this particularly in relation to PEG feeding. However it was capacity issues on a day-to-day basis which were shown by this study to be particularly difficult for the family carers and staff who had to live with, or contest, these decisions (Wilson et al. 2010b). As illustrated in Chapter Seven, decisions are often made in a person's best interests despite them having some level of capacity, particularly in relation to balancing risk and independence. The third of the five key principles of the Mental Capacity Act addresses unwise decisions such as refusing to undertake personal care, to eat properly or engaging in risky behaviour. This principle sets out that just because someone's decision could be considered unwise does not mean they lack capacity to make that decision. However, for people with HD capacity may be considered on a number of levels or it may fluctuate, making it a challenge for others to know when capacity is sufficiently impaired to require decisions being made by others in their 'best interests' (Department of Constitutional

Affairs 2007). The Mental Capacity Act also does not consider the rights and interests of the person caring for the patient with HD.

In this study I observed decisions being made on a regular basis. These seemed to be instigated through collaboration with health professionals, family carers and the person with HD. Hence I draw on the concept of relational autonomy as described by Ho (2008, also see Nolan et al. 2004, Lloyd et al. 2012, Verkerk 2001). In this context relational autonomy encourages us to view the patient as more than an independent unit and recognises that patients should be situated within 'a rich matrix of relationships and socio-cultural beliefs' (Nolan et al. 2004, p47). From this perspective, we accept that decisions about care are not just related to the illness but the social, psychological, and historical nature of living with the condition and as such patients and families live 'intertwined lives' (Ho 2008, p132).

Ho suggests that contemporary medicine and its specialism's have inadvertently reduced patients to 'diseased body parts and medical jargons' (2008, p130). It is such depersonalised and compartmentalised care that I argue is unsuitable for those with HD as the bigger picture of their lives and relational identities are so essential to quality care and appropriate decision making. The decision making I observed and that was reported to me was very much approached in this way. The key health and social care professionals had built knowledge and an overarching view of the person with HD and their family situation so that decisions could take into account more than just the biological needs of the person with HD. Therefore I suggest an expansion to the notion of relational autonomy to include the health and social care professionals as legitimate participants once a relationship of trust is built.

This study has shown that a particular challenge arose when the wishes of the person with HD conflicted with the wishes or needs of the family carers. Amy and her husband Ben provided a clear example highlighting the issue of whose autonomy and 'best interests' were being made a priority. It is apparent that a patient should not be forced to do something that they do not want to do but we should query how much decision making ability may be altered, and whether

that should have an impact on the validity of the decision. This presents professionals and families with complex situations for which there do not always seem to be appropriate or 'best' solutions. In this study, Amy's autonomy was accorded precedence over Ben's needs, yet in the longer term this may have ultimately jeopardised her wishes.

To identify when capacity becomes an issue is a challenge for families and health professionals. In this study family carers grappled with a balance between respecting their loved one's autonomy and fearing they were unable to make appropriate decisions. Slowly diminishing capacity presents a particular problem for decision making in terms of balancing these practical and ethical issues. For those caring for someone with HD the challenge is to know whether behaviours are a symptom of the condition or a 'choice'. Whether or not this affects the decision to be made was then an issue of debate for both families and health and social care professionals.

I suggest that a more realistic approach to family carers' needs should be taken. There is increasing recognition of the financial and labour costs of caring and that these could not be met without family caregivers (Carers UK 2004, Buckner and Yeandle 2007). Perhaps there needs to be a wider recognition that it is family carers that need to be maintained to allow a person with HD to live at home for extended periods. Adopting a relational approach demonstrates that it is necessary and reasonable to consider, and potentially prioritise, the interests of family carers in a more direct way. This entitlement would place the family carer's interests as equally important, thus 'intertwined' (Ho 2008, p132) with those of the patient.

Health professionals need to begin negotiation and guidance with the person with HD and their family carer early in the disease process. Evidence from this study shows that a built knowledge and rapport with families through continuity of personnel engenders trust in health professionals, and this trust is essential in creating a more conducive environment in which important decisions can be made. When the person with HD, their family and the health and social professionals knew each other and had a built trust more cohesive

decisions were made. For example, one patient participant in this study wished to draw up legal documentation regarding his wishes for care. In this instance the care home requested a GP and a psychologist to assess his ability to make these decisions. Again, however, it was the long-term relationship with the CNS that was called upon to assist with validating these decisions and to assess whether they had changed over time. Family carers also talked of using health professionals to initiate conversations about difficult decisions such as PEG feeding. This suggests that families consider staff to be part of their relational system particularly when it comes to decision making.

Personal autonomy and uninfluenced decision making is a dominant rhetoric in health services. This study shows that in reality decisions are not made in isolation but in collaboration with, not only family, but also trusted health and social care professionals. This suggests that a broader version of relational autonomy is more appropriate for those with HD. Directed decision making provides people with HD and their families with support for decisions and allows for appropriate care planning within the constraints of resource limitations.

HEALTH AND SOCIAL CARE PROFESSIONALS

Health and social care professionals talked about building trust with the person with HD and their family. In the early stages professionals made efforts to balance being available for contact and over surveillance in order to start building a good rapport. Some services were introduced in the early stages, prior to need, not only to help maintain function but to normalise the service input and reduce anxiety when further input may be needed. Several patients and family carers recounted stories of difficult encounters with professionals who knew little of the disease or their family life, particularly those making assessments for needs and benefits. In these situations families turned to the health and social care professionals they trusted who not only understood the complexities of the condition, but also knew them as people.

The stability of the services and the individuals within key roles was clearly a factor in building trust. In this study all the key health and social care professionals, with the exception of one consultant, remained in their roles throughout the four year data collection period and had been in those roles several years prior to the start of the study. Larch Tree care homes also reported low staff turnover. It could be suggested that this level of permanence within posts may be distinctive and may not always be possible. Although by no means conclusive, this study indicates that HD services attract a certain type of person and that once involved in caring for people with HD health and social care professionals continue to work in this field.

SERVICE PROVISION

Health care models are often used to illustrate key features that service providers should strive for when planning health services. Health service planning has previously been based on the need to provide rapid responses to acute illness or injury. The rise in incidences of chronic conditions has prompted a shift in types of care and the ways in which care is provided. The chronic care model suggests a change in the organisation of care for those with chronic conditions (Wagner et al. 2001). While care models such as the chronic care model (Wagner 1998) can have some application to HD, flexibility and caution must be applied. The findings of this study and previous research have shown that the central element of a model of care for HD (Figure 11) is to take a holistic approach and involve a multidisciplinary and multifaceted team (Veenhuizen and Tibben 2009). I suggest that surrounding this premise a key worker approach should be utilised and continuity of care maximised across all aspects of service provision. Ongoing work by NHS Improvement (2012) teams is currently focusing on providing equitable care seven day a week. An important element is the accessibility of care. Such equitable access challenges the desire for continuity of care so important to those affected by HD, creating an additional balance for care providers to strive to achieve.

The key worker develops disease, patient and family specific knowledge and a wider knowledge of local services. This is a single point of contact and co-

ordinator of care services for families promoting continuity of service and personnel, something specifically important to those with HD. The key worker role is therefore illustrated as part of the triad of care in addition to the person with HD and the family carer. A model of care for HD should promote ways for people to manage by themselves for as long as possible and create services that are accessible when needed. Where they are available and willing to provide care family carers are fundamental to the process. A family carer will often take on the role of co-ordination of, and liaison with, services. As cognitive function decreases the family carer will take over the majority of organisational tasks alongside the physically demanding role of caring.

The model below is based on the current literature, UK policy and evidence from this study and may be a useful summary for the development of long-term care provision. The model shows how symptoms increase over time as does the need for service input. I have suggested the types of services that might need to be introduced over the phases of HD. However, both the type and timing of such services should only be considered as a guide. Not all people with HD will require or want these services. This study has shown that patients and families dip in and out of services throughout the course of their illness, with the appropriate services being provided as and when needed. Moreover, this study has highlighted that consistency in services and personnel was of benefit to those with HD. Not only because those with the condition find change particularly disruptive, but also because it allowed staff to build the disease, person and service specific knowledge necessary for quality care.

The model also shows the potential tipping point at which time it may no longer be possible to care for the person with HD at home. This study has shown that for those without family carers this may occur earlier in the illness trajectory. At this stage the balance between care provision and need may be tipped and a move to residential care may be necessary. Respite, day care and home care services may also be needed throughout the illness trajectory. Again, what type of service and when it is needed will vary depending on the patient and their family. As a valued member of the triad of care the key worker should take a role in organising and co-ordinating all care input. The specific application of

the key worker role, specialist knowledge, continuity of care and self-management are discussed later in this chapter.

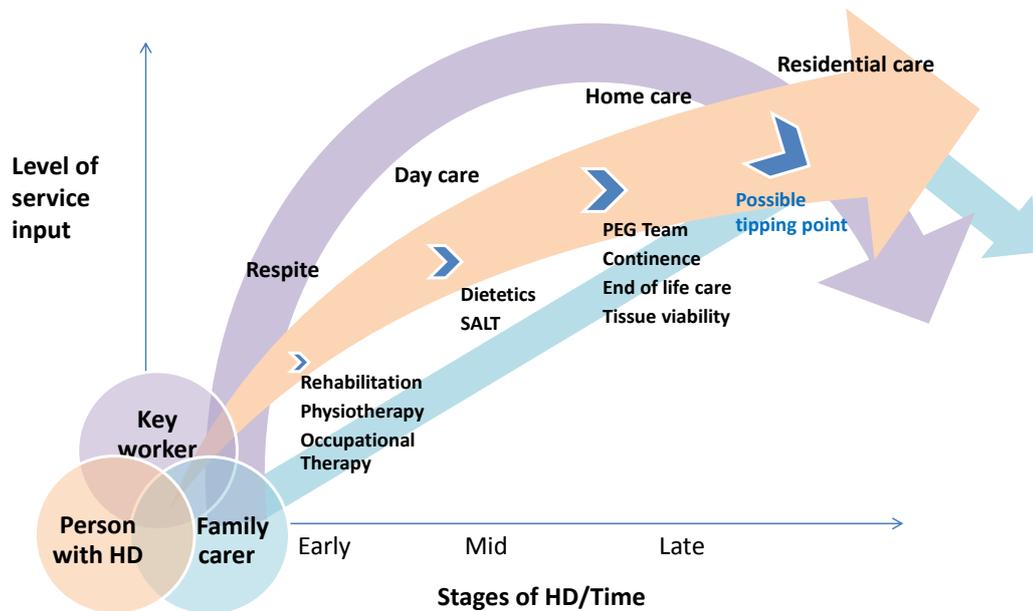


Figure 11 – A model of care for HD

Throughout the disease trajectory health and social care professionals are involved in the care of people affected by HD. This care may be directed at the person with HD, their family carer or wider family members. In the early stages, health and social care participants in this study reported regular input with both patients and families. This type of care focused on helping them to adapt and cope with a new diagnosis as well as providing information. As families adjusted input was often reduced to a status of monitoring via out-patient clinics, telephone contact and home visits. This monitoring was to allow health and social care professionals to react quickly to changes that occurred and to try to prevent crisis. This type of approach to HD services is supported by work conducted by Simpson (2007) who advocates a shift in focus from crisis intervention towards models of chronic care.

During the mid stages of the disease the input from health and social care professionals was more focused on maintaining health and well being and co-ordinating the increasing numbers of additional services that needed to be instigated. At this stage the professionals' knowledge of the condition, helped with symptom management, and links to a wider multidisciplinary team became essential. A supportive, informational and advisory role was also maintained.

As the disease progressed into the later stages, professional input intensified and availability of a central contact became vital to co-ordinate the myriad of services involved in providing complex care. If the person with HD remained at home, co-ordination of day care and respite services was important to support family carers in maintaining the balance between care input and need. When sustaining the person at home was no longer possible, a move to residential care occurred. All of the residential care homes in this study had experience and knowledge of HD (Smith 1998). However, when necessary they drew on the advanced expertise of the clinical nurse specialists. The CNS also continued to provide a link to wider service resources and medical knowledge, even after the patient had moved from the community to residential care. The link with the CNS also remained an important constant for the family carers. Through this continued relationship the CNS provided support, information and contact between the care homes and community services such as neurologists. They also provided education and support to care home staff.

Both the Birchtree and Oakfield community services could be considered positive examples of the multi-disciplinary care advocated by policy and research for those affected by HD (Clough and Blockley 2004, Department of Health 2005, Skirton and Glendinning 1997, Oliver and Borasio 2004, Veenhuizen and Tibben 2009, Aubeeluck and Wilson 2008, Kenny and Wilson 2012, Royal College of Physicians 2011). In Birchtree the service was led and co-ordinated by the CNS but the HD team included a consultant, speech and language therapist and dietician on a secondment basis and further referrals to wider services such as occupational therapy, physiotherapy and psychiatry were also made. In Oakfield, the service was provided by the consultant, supported by the RCA for the area whose position was funded by the

Huntington's Disease Association charity. Wider referrals were made to all the relevant supporting services. These community based services also demonstrated their role in the provision of expertise and advice to residential care homes. The residential care homes in this study had high levels of experience in providing care for those with advanced HD. However they also relied on their links with community based staff for multidisciplinary input such as speech and language therapy, and specialist support from neurologists and clinical nurse specialists. All three types of services demonstrated that even when the case-load does not warrant a full time multidisciplinary team, secondments, third sector contribution and good referral links can help maintain a quality service. The advantage of the use of secondments is that the same professionals are involved each time, building their expertise and rapport with families.

KEY WORKER

A significant component highlighted by the findings of this research has been the importance of the CNS and RCA roles as key workers in community care. These personnel provided a single point of contact for people with HD, their families and other professionals including care homes (Royal College of Physicians 2011). They acted as advocates, liaisons and care co-ordinators working across disciplines, organisation and sector boundaries. Aspinall et al. (2012) identified the clinical nurse specialist as best placed for this role of case manager navigating across and between services. A single point of contact and open referral system allowed easy access for patients and families amidst a confusing array of appointments, different health professionals and supportive services. The Royal College of Physicians (2011) also advocate for a key worker role for the care of those with long-term conditions. The report recommends specialist nurses to be central to delivery of quality care alongside GPs with a Special Interest in specific conditions.

This central role is referred to in a number of different ways such as care managers, case managers, link nurses, discharge co-ordinators, or a liaison nurse. I propose the term key worker is most transparent and appropriate for

the care of those affected by HD. As Payne et al.'s (2002) review of the literature on the communication of information about older people between health and social care practitioners demonstrates, there is little consensus about who can be a key worker or the specific elements of their roles. Historically, these roles have been taken on by social workers (Challis 1990). However as Clausen et al. (2005) note, time and case-load constraints for social workers have reduced their ability to work in the traditional 'casework' role with individuals. The trend is now toward social work involvement only when there is a specific need rather than a comprehensive holistic approach for a continued period of time. The authors suggest that the non-medicalised, 'jack of all trades' aspects of the role allow social workers to operate proactively in advocating for the patient and family (2005, p280). Evidence from this study suggests that it was precisely this broad non-medicalised role that was problematic for those needing help when affected by HD. Participants in this study praised the specialist knowledge of the CNS and RCA and appreciated their links to other specialist health and social care services.

The key worker role has been in place in the community for some time in various forms with application to mental health, learning difficulties, and older people in the community (Payne et al. 2002, Challis and Davies 1985, Dant and Gearing 1990). The main application of the role has been to facilitate co-ordinated care of chronic illness and disability in children (Rahi et al. 2004, Greco and Sloper 2004). The paper written by Greco and Sloper (2004) describes many transferable aspects of the key worker role. The authors highlight the input received by disabled children and their families from a variety of different services such as health care professionals, social work, education, benefits and voluntary agencies. They suggest the role of the key worker is therefore to 'represent the single point of contact that families would like' (2004, p13). Taking a quantitative questionnaire survey approach, the study identifies some areas of good practice such as open referral, allowing families to refer themselves to the scheme, and planning meetings with families to avoid duplication or omission of services and support. These are all aspects that have been illustrated as important in this study and are described by

Veenhuizen and Tibben (2009) as part of an out-patient clinic approach to co-ordinate care for people with HD.

Work commissioned by the Welsh Office for Research and Development provides a review of key worker systems for children with disabilities (Liabo et al. 2001). The key messages from that report strongly supported the key worker approach, finding that when a key worker was involved the overall quality of life of families was improved, relationships with services were better, access to benefits was broader and quicker and levels of stress for the family were reduced. Parents also reported a good relationship with the key worker as an important factor in itself. It is of note that no evidence was found that the key worker system resulted in variations in quality of medical care. Although in the field of elderly care, Payne et al.'s (2002) systematic review found that the most effective strategy for transferring information between health professionals was the use of key workers to provide a point of contact for professionals from hospital to the community. Interestingly this paper does not address how key workers might improve information to patients and or families, but focuses on communication between professionals across service boundaries. Throughout the variety of papers reviewed by Payne et al. (2002) key workers were considered important in improved discharge planning and co-ordination of services both at hospital and in the community. This was demonstrated even in non-complex cases where the key worker was not directly involved. This transcendence of professional, organisational and sector boundaries is essential to facilitate communication, co-ordination and co-operation between the many services people have to interact with when they are affected by HD.

In this study the CNS and RCA took on this key worker role by providing access to and co-ordinating services for families, as well as offering support. They built professional yet friendly relationships with patients and families and were considered a 'first port of call' for help and advice. In the participating care homes residents were allocated a key nurse and a key support worker to specifically work with that person, advocate for their needs and act as a point of contact for other services and family members. Those operating in such key worker roles have reported high levels of satisfaction with the role (Liabo et al.

2001). Such satisfaction may go some way to explain the long-term commitment to services that professionals' in this study demonstrated.

SPECIALIST KNOWLEDGE

Several elements made up the skills employed by the CNS and RCA. Participants in this study also noted personality traits that aided their rapport with these staff. However the fundamental aspect was considered to be their knowledge base. This included their specialist knowledge of HD, their built knowledge of the person with HD and with their family, and the wider knowledge of health and social care systems such as benefits, local centres for day care and respite and links to other services such as physiotherapy, occupational therapy, speech and language therapy and dietetics. Where possible, it was seen as beneficial for professionals in these wider services to also have such a knowledge base. As the staff participants in this study noted, they felt they were able to provide a better service to patients and families because of their knowledge and experience of HD which they would apply to the knowledge they developed of each family.

Liaschenko and Fisher (2004) suggest that nursing knowledge is made up of case knowledge, patient knowledge and person knowledge. The authors suggest that the work of nurses goes far beyond that of the biomedical knowledge necessary. They suggest there is little recognition of the co-ordination of care that routinely takes place and is informed by this range of knowledge types (Liaschenko and Fisher 2004). I argue that it is not necessarily the nursing element that has been shown to be essential in HD service provision but the HD specific knowledge. Hence families reported that the disease specific knowledge was important for all health and social care professionals with which they engaged.

According to Liaschenko and Fisher (2004) 'case knowledge' is the disembodied disease specific biomedical knowledge which is often not found outside specialist services for HD. It is this case knowledge that gives staff cues as to what can be expected and is therefore essential for quality care planning. Liaschenko and Fisher (2004) suggest this is the most straightforward and easiest knowledge to work with. This study has however highlighted that for

those working with people affected by HD this biomedical knowledge of the disease is limited and therefore highly valued by patients and family members. The health and social care professionals with this specific case knowledge were often used to filter this information to other professionals and families. However there were also clashes with care providers who did not have this case knowledge and did not understand the implication of the condition, in particular the recognition of both physical and mental aspects. Families and the staff in this study reported instances (see 'Funding and Bureaucracy' in Chapter Seven) when inappropriate assessments were made for funding and care needs due to a lack of understanding of the condition by those in wider roles such as Social Services and assessment teams.

In this study the CNS in particular took on all of the aspects of what is termed 'patient knowledge' (Liaschenko and Fisher 2004) by negotiating between services, across organisational boundaries and 'marshalling appropriate resources' (2004, p47). Patient knowledge is seen as how a person becomes created as a patient by gathering certain information about them, how to get things done for that patient, across and within organisations and knowledge of the others who might be involved in that patient's care. From extracts in this study I have shown how the CNS was involved in helping to co-ordinate care services such as day care, respite and residential care and referrals to wider services. Nurses also took a key role in liaising between families, the residential care home staff and the consultant. Residential care home staff talked of bringing in additional services when needed to meet the changing care needs of residents.

It is clear from this study that 'person knowledge' was built up by a range of health and social care professionals over a number of years. Liaschenko and Fisher (2004) identify person knowledge as knowledge of the person beyond that of their biomedical self. This type of knowledge engages with the person's history, spatial and social situations. The authors suggest this can only happen over time over several encounters or during an extended period. For staff in this study the person's history was an essential part of this and fed directly into the concept of relational autonomy. During consultations genetic lines of inheritance were

mapped to identify those at risk of the condition. Home visits allowed the health and social care professionals to acquire knowledge about spatial and social circumstances in what Bjorkland refers to as 'invisible triage' (2004, p111). Staff at residential care homes also gathered this person knowledge. These staff considered 'getting to know the resident' vital to providing tailored, personalised, and quality care (Wilson et al. 2011).

I would argue that any key worker for HD would need all three of these types of knowledge to provide effective care and case manage those affected by HD. Work by Rasin and Kautz (2007) has shown how these different types of knowledge become essential in dementia care in order to manage behaviours yet keep in mind the needs of the person exhibiting that behaviour.

CONTINUITY OF CARE

Continuity of care, especially within the key worker role was also an important element of care for those participating in this study. Findings in this study show the importance of building relationships between those affected by HD and the key health and social care professionals they came into contact with. For the professionals, getting to know the patient and their family was an important tool in assessing the best way to balance level of input, manage care, deal with crisis and plan future decisions. For the person with HD and their family carers, building trust with the health professional allowed them to respect their advice, engage with them for decision making, feel comfortable to ask for help and be happy with them in their home.

Continuity of care is taking on increasing importance in the fields of dementia and the care of older people (Cornwall et al. 2012). Freeman and Hughes (2010) suggest that continuity can be broader than the expectation of dealing with the same health professional over time. They suggest both relationship continuity and management continuity. Relationship continuity is defined as 'a continuous caring relationship with a clinician' and management continuity as 'continuity of clinical management, including providing and sharing information and care planning, and co-ordination of care' (p13). Aspinal et al. (2012) add personal agency to this list through their systematic review of the literature. These

different elements of continuity focus on care which provides; one or more named individuals with whom the person affected by HD can establish and maintain a therapeutic relationship, uninterrupted care for as long as is required, a smooth transition between care providers, adjustment to the person's needs over time, transfer of information, ways to sustain the person's social and personal relationships in the community and enhance quality of life, scope for people to retain control over their own lives and to manage their own health and well being (Aspinal et al. 2012).

This study has shown that continuity of staffing makes a considerable contribution to knowledge of the patient and the family. This knowledge, gained over time and through repeat interactions allowed health and social care professionals in this study to have a better understanding of patients' wishes for care, their capacity to make decisions, and the types of decisions they were able to make. This was viewed in context of the patient's social, spiritual and relational (including family carers) circumstances. The longitudinal nature of this study and the longevity of the condition means that participants were able to describe, and I was able to observe, the importance of such continuity.

However, this study also raised issues of accessibility and busyness of health and social care professionals. All patient and family carer participants noted the perceived availability of the CNS and RCA but also expressed their reticence to access them due to their busyness. Aspects of entitlement and perceptions of personal versus other people's need were also elements causing patients and family carers to self ration their use of the key resources such as the CNS and RCA. It could also be queried whether this was also partially an expression of independence. By not contacting services unless necessary patients and families generated self-efficacy by managing by themselves. The built relationship between those acting as key workers and family carers may also permit family carers to associate the key worker role with what they do as carers. In this way they understand and respect nature of the role and may be seen as including the interests of the health and social care professional in their relational autonomy. Hence part of the reciprocity of the triad of care (patient, family carer and key worker) is therefore, to ration the use of resources.

A key issue for HD services is how to achieve continuity of care without creating dependence on one specialist member of staff. The Birchtree team recognised such dependence could be problematic and introduced a further member of the team to take on some of the work of the CNS and create an additional point of contact for those in the HD community. In Oakfield there was considerable uncertainty as the consultant was nearing retirement age and it was not clear who might take over the running of the HD service. The RCA was a charity funded post already stretched to cover a large geographic region making the service in the area somewhat unstable. The availability of the HD team in Birchtree allowed the RCA to concentrate her time in and around the Oakfield area. Such reliance on the third sector to provide care is set to increase as the government brings about changes to widen the role of the voluntary sector and charities in health care provision (Department of Health 2011).

Indeed a report by The Centre for Social Justice recommends that the third sector take on roles of care co-ordinators or key workers for those with mental health issues (2011). The report suggests embracing the Department of Health's 'Any Qualified Provider' scheme by using voluntary services and charities to fulfil this role of care co-ordinator. The report argues that as NHS Trusts and Social Services provide the majority of services, this creates a conflict of interests when giving information to, and advocating for, patients and their families. I have previously highlighted the benefits of health and social care professionals directing patients and families in order for them to make decisions about their wishes and needs. Third sector staff could be used as independent key workers to provide information and advice that is less driven by the financial restrictions of the NHS. Alternatively, it may be that they could be less informed about the restrictions under which services must work. The report recognised the need for high quality 'skills, sensitivity, experience and awareness of services available locally (across all sectors)' (The Centre for Social Justice 2011, p218). It is not clear how this would then allow third sector employees to maintain independence from financial restrictions on resources. However patients and families are supported, there will always be a need to balance wishes and needs with the availability of resources and costs. In this

study there was a clear reliance on the Regional Care Advisors provided by the Huntington's Disease Association. Although stretched this input was working well to support the out-patient clinic service in the Oakfield area. Yet this link with the NHS service had been built over many years between a specific RCA and a consultant with an interest in HD. The loss of one of these people may significantly change the dynamic and delivery of the service. If a reliance on the third sector is to increase, there will need to be increased funding and support for these personnel. Their roles will also need to be given due consideration and weight when advocating for patients and providing information to professionals.

SELF-MANAGEMENT

Evidence from this study shows that it is achievable for patients and families to manage by themselves, to a point. I have argued that even for those in the early stages when intervention for physical tasks may not be necessary the health and social care professionals maintained regular input beyond that of annual clinic appointments. This input was primarily to provide support and information. While one participant in this study requested time away from health care to come to terms with their diagnosis, others reported regular contact with the CNS via phone and home visits.

Self-management can mean different things to different people and not all will want to, or be able to participate in self-management techniques or formal programmes. In a report commissioned by the King's Fund Corben and Rosen suggest that to most people self-management means 'developing an understanding of how their condition affects their lives and how to cope with their symptoms' (2005, vii). Whether or not people want to, and do, engage with self-management will be dependent on a number of aspects including time since diagnosis, severity of the symptoms, age, level of education and their social, psychological and emotional needs (Corben and Rosen 2005).

Two studies have focused on formal self-management programmes for HD. Campo et al. (2012) found that an educational intervention programme

supported self-management strategies and improved psychological well-being and coping strategies for both patients and family carers. The programme sessions included education and discussion on information seeking, taking a pro-active role in treatment, self-monitoring of body, cognition and mood, relaxation and stress management by focusing on realistic and helpful thoughts, dealing with and preventing depression, social competence and asking for social support (Campo et al. 2012). The findings showed evidence that the programme was less effective for those in the pre-manifest stage of HD. Although those with pre-manifest HD and their family carers used more seeking of social support after the programme they did not have any improvement in the psychological outcome measures taken directly after the programme (Campo et al. 2012). However, there was no longer term follow-up or assessment of the economic feasibility of the programme.

In contrast a randomised controlled trial for people with progressive long-term conditions, including HD found that an education programme delivered by an occupational therapist to reduce the risk of falls and pressure sores had negative effects (Ward et al. 2004). Those randomised to the education programme arm of the study were comparable at baseline with those in the control group. Post programme outcomes for those in the intervention group at 12 month follow-up showed declining functional ability, with no positive effects on well-being, falls or skin sores (Ward et al. 2004). This research by Ward et al. (2004) challenges the assumption that education and self-management interventions are beneficial and supports the need for further empirical evidence of their effectiveness, particularly for those with degenerative conditions.

Participants in this study engaged with many forms of what could be termed 'self-management' techniques such as adapting their behaviour to sit down when they got dressed, taking exercise to stay physically active and doing crosswords to sustain mental acuity (Chaplin et al. 2012). It could be argued that throughout the reported findings are examples of health and social professionals providing one-to-one 'self-management' advice on all the elements identified in Campo et al.'s (2012) programme.

All staff participants provided patients and families with information and techniques to maintain health and well being. The rehabilitation support worker specifically worked with patients to improve and maintain their physical abilities. I suggest that for those with HD this type of approach to helping people manage by themselves may be more beneficial than formal programmes.

It is clear from this research that where possible any advice or programme to promote independence from services should embrace family carers (where available), not only to assist the person with HD, but to encourage carers to initiate ways to maintain their own health and well-being. Participating family carers reported the benefits of a more formal information and support day provide by the HD team in Birchtree. Further research is needed to ascertain whether more formal programmes may be more appropriately aimed at family carers rather than patients. Continued shame and silence associated with a diagnosis of HD may prevent some from attending such days. For patients, as with clinic appointments, mixing those in the early and later stages may be stressful for some (Chaplin et al. 2012). Due to the small numbers of families affected by HD separating into groups according to disease stage may not be feasible. However, in line with the theory behind current policy it could be queried whether group education and information programmes could reduce utilisation of services (Gately et al. 2007) such as the number of home visits provided by the CNS.

Some form of support to manage by themselves, particularly in the earlier stages of the disease may be desirable and appropriate and a number of patients and families made efforts to engage with this. However, this must be seen in conjunction with open referrals and easy access to specialist care when needed. Any form of 'self-management' can only work proficiently when patients are given information and support to manage their condition and know when it is appropriate and necessary to seek additional input from health professionals (Chaplin et al. 2012). This study has illustrated that for those with HD it is not only physical ability to

perform personal care and tasks of daily living that must be considered. The person's cognitive functioning should also be a key factor in whether they can manage these tasks. When symptoms such as apathy and lethargy are prominent managing on their own may not be appropriate. As the health professionals in this study noted they are not available 24 hours a day and rely on family carers to make assessments, keep the professionals up-to-date with accurate information and contact them for help when needed. When family caregivers are not available and the person with HD lacks the self awareness to monitor their own health things can deteriorate to a crisis situation. A report by the Centre for Social Justice draws on several initiatives such as 'Shared Lives', which aims to create family and community orientated living situations to keep people in the community (The Centre for Social Justice 2011). It could be possible to extend these types of mental health based schemes to those without family support living with the early stages of HD.

There is a need to look critically at how appropriate self-management programmes may be for people with HD in light of potential declines in cognitive functioning. There is no other research exploring education programmes, self-management or what it is like to live with HD in the early stages beyond the period of diagnosis. Hence there is no evidence of the validity of the self-management approach for HD over long periods or how this might be sustained and delivered alongside services. It is therefore not possible to assess the efficacy of programmes using evidence derived from those designed for Parkinson's disease, multiple sclerosis or other long-term conditions (Rae-Grant et al. 2011, Macht et al. 2007, Chaplin et al. 2012). Helping people to manage by themselves and formal self-management for HD therefore requires individual consideration. This is a significant gap in the knowledge base which should be addressed to inform the types of support and the ways in which those with HD in the early stages and their families want to be helped to manage by themselves.

CHAPTER TEN: CONCLUSION

This is the first longitudinal qualitative study of the experiences of those affected by HD. Fifteen case studies were followed for a three-year study period. Findings from this study are consistent with the existing literature on service provision, particularly in regard to multidisciplinary input. However, a number of insights into living with HD and how services might better provide for such a complex and multifaceted illness have been highlighted. This chapter concludes this thesis by drawing together these areas of discussion.

People with HD can be affected by motor, cognitive and behavioural symptoms and it is this triad of symptoms coupled with the genetic nature that makes the disease so complex requiring multifaceted and multidisciplinary input. The condition has far reaching implications for biological family members who may be at risk and heavy burdens are placed on those caring for a family member with HD. The person with HD and their family adapt to cope with a long-term progressive condition in order to normalise their lives. Slowly diminishing capacity and behavioural issues must be balanced with striving to maintain independence and autonomy.

The Global Burden of Disease survey (Menken et al. 2000) has identified neurological conditions as an increasing issue for health services. In the UK the National Service Framework for Long-Term Conditions was rolled out in 2005 but there have been concerns that targets are not being met despite large amounts of money being invested. This is thought to be due to a lack of leadership and infrastructure, such as a lack of neurologists. There is some policy guidance for the UK but little is specific to long-term neurological conditions and there is no national guidance for HD. Information on managing the condition is predominantly provided by the national charity for the condition, the Huntington's Disease Association. In response to the lack of improvement since the implementation of the National Service Framework the report by the Royal College of Physicians (2011) made a number of recommendations for service improvements for those with long-term conditions including the use of specialist nurses, GPs with a Special Interest, key

worker roles, clear and unrestricted links to neurological services and a more cooperative approach between neurology and psychiatry.

Research on HD is primarily focused on the search for a cure and much is being done to better understand the way in which the mutated gene affects the brain. Policy and evidence from research focused on care delivery supports a multidisciplinary approach to care, although there have been few studies that evaluate services and little is known about what it is like to live with HD. Self-management is encouraged for those with long-term conditions however there is little consistency in meaning. There is no evidence of the validity of the approach for HD over long periods or how this might be sustained and delivered alongside services.

This study used longitudinal case studies with the person with HD as the central component and included family members and health and social care professionals. Ontological stance provides that HD be viewed as more than an isolated biological disease. The context of all aspects of the person, their relational, social, psychological and historical systems are also taken into account. In order to understand living with HD from this wider perspective this unique study used multiple methods and multiple sources to build a picture over an extended period of time.

Methods of data collection included the responsive interviewing technique alongside observation, eco-mapping, and genograms. These were used to gather data from the person with HD, their family carer and a nominated health or social care professional to build fifteen cases involving 33 individuals and 115 interactions (68 interviews and 47 observations) over three years of study participation.

This approach recognised the role of all participants including the researcher in the co-construction of the data. Considerable efforts were made to maintain a flexible approach to data collection in order to accommodate the physical, social and time constraints of patients, families and the health and social care professionals. An adapted information sheet and consent form was used for recruitment. Continued learning from these experiences required

methodological adaptations, particularly to interview techniques. Analysis was undertaken to interrogate the data both within and across cases. Findings were presented to give context to each case (Chapter Four: Building cases and Chapter Five: Case profiles) and then a temporal approach was applied to the data to examine themes across the fifteen cases (Chapters Six, Seven and Eight).

Findings show that living while affected by HD requires continued readjustment to maintain balance between increasing disability, diminishing cognition and living well at home. This ongoing modification is undertaken by patients, their families and the health and social care professionals that provide services and support. This study has explored the complexity of living with HD, the wider genetic implications, and the nature of providing care for someone with HD. It specifically adds new dimensions to what is known about living with HD by incorporating multiple perspectives over time. It is this concept of balance that has arisen throughout the findings and has been used to try to encapsulate the continued negotiations that families and health and social care professionals must undertake to try to maintain the delicate equilibrium of living with HD.

Patients and families are challenged with coming to terms with a multifaceted condition which affects them physically, cognitively, psychologically, behaviourally, socially as well as having broader genetic implications for the next generation. Families can face dramatic changes to their circumstances and may experience increasing encroachments on their home as the disease progresses. Chronic and long-term illnesses can be seen as disruptions to a person's biography and sense of self. Evidence from this study shows that normalisation can occur after a period of initial adjustment. This can be sustained through constant realignment until the balance of care needs and provision is finally tipped in the more advanced stages of the disease. Once care needs can no longer be met in the family home environment a move to residential care is necessary. This move can send family carers into a liminal state when they must shift from an active and physical 'carer' role to a more passive 'caring' one. This study has shown how family carers struggled to hand over care to professionals and move forward to re-establish themselves outside the realm of caring and HD.

HD does not fit with the current compartmentalised, depersonalised health and social care system. This study has highlighted the role of the multidisciplinary team as recommended by current policy and the literature. However, case knowledge of the disease, patient knowledge of the health and social care systems, and person knowledge of the patient and family becomes essential to providing appropriate care. Participants demonstrated the importance of a key worker role in co-ordinating care, identifying future needs, decision making and maintaining autonomy. A personalised and holistic approach to care is therefore essential.

The concept of relational autonomy has been used in this thesis to illustrate the context in which people with HD live. Their decisions are not often exclusive to themselves and reflect their social, relational, environmental and historical circumstances. I have argued that this relational system is made up of a triad of key participants in care; the patient, the family carer (where available) and a key worker. Health and social care professionals must balance the best interests of the patient, the family and the limited resources with which they work. Decisions are therefore necessarily steered or directed by health and social care professionals. This is not necessarily negative and can alleviate some of the considerable burden of decision making often placed on people with HD and their family carers.

This study has highlighted that fluctuating, and slowing diminishing capacity and cognitive decline in HD poses challenges for families and health professionals. In these instances the balance between autonomy and risk is delicate and complicated, particularly for family members. Staff knowledge of the patient and their wider circumstances can significantly help them to support families with challenging decisions and care planning.

The fundamental issue for patients and families was to balance the burden of care with the input of necessary services. Four key aspects of this balance have been identified:

- The management of knowledge and information: the burden of genetic information necessitates a balance between sharing and informing with secrecy and silence.
- Autonomy, choice and decision making: balancing autonomy with best interests and whose choices and decisions are given priority.
- The transformation of homes to hospitals: balancing the home environment with the intrusion of necessary services and equipment.
- Burden of care is shifted but not lifted by a move to a residential care home: balancing being involved with the person's care and moving on to create a life as an individual.

By including health and social care professionals in this study it has been possible to see how these issues for patients and families have been addressed. Local out-patient services and residential care homes used four key elements to help maintain balance:

- Multidisciplinary working
- Key worker approach
- Disease, person and service specific knowledge
- Continuity of staffing and care.

These aspects of care were able to promote living independently to an extent in the earlier stages, particularly when family carers were available.

Multidisciplinary working allowed access to a range of extended services such as speech and language therapy, dietetics, and physiotherapy, all of which were considered important for improving or maintaining health and well-being.

Generalists' limited knowledge meant that the disease specific knowledge of the specialist team members was highly valued. The continuity of staffing within the teams and within key roles promoted trust and aided decision making and autonomy. The key worker approach allowed for one point of contact for access to wider services, which supported continuity, trust, and information sharing. This person also developed an overarching view of the patient and their family situation in order to inform care planning and decision making.

However, this study has illustrated several challenges for care providers. Services recognised that one person with specialist knowledge working in a key worker role created dependence on that person. Services were challenged to provide care when that person was not available such as during annual leave periods. The Birchtree team had sufficient funding and population of patients to add an additional part-time team member to take on some aspects of this role. It must be recognised that this may not be feasible in all NHS Trusts. Rarer conditions such as HD pose challenges for equity of service provision. Smaller pools of patients may not warrant specialist nurses or multidisciplinary teams but improved national networks and co-ordination of services are needed to make sure access to services is not dependent on where people live. If the recommendations set forth by the Royal College of Physicians (2011) are to be implemented for HD attention needs to be paid to the national lack of neurologists in the UK and the division between neurological and psychiatric services. In this study psychiatric input was considered extremely difficult to attain for HD patients with services often not recognising both the mental and physical aspects of the condition.

A further difficulty for the health and social care professionals was to maintain the health and well-being of the family carer. Staff recognised the importance of the family carer in the care of the patient at home but were sometimes challenged to privilege the patient's wishes over the needs of the family carer. Family carers often played a key part in patients managing without the input of services. Health and social care professionals relied on family carers to provide some form of surveillance and alert them to problems as they arose. Health and social care professionals were therefore challenged to find the best ways to promote and sustain people to manage by themselves whilst still maintaining contact in order to plan care and avoid crisis interventions.

Finally health and social care professionals reported on the challenges of liaising and working with other professionals who lacked knowledge about the complexity of the condition, particularly those making assessments for funding and equipment. They were therefore challenged to find effective ways to up-skill generalists by sharing information and providing education. In this study

this was particularly evident in the good relationships built between community-based staff and the residential care homes.

AREAS OF FURTHER EXAMINATION

This study has explored the HD services offered in two regions of England and those provided by a national charity running a number of neurological care centres across the UK. This investigation has highlighted the difficulties in identifying the location, type, and availability of services elsewhere across the country. A national scoping exercise is now necessary to establish how services are being delivered UK wide. This study has shown that services can be provided in a variety of ways and these should be tailored to the population of people with HD, their needs and the health and social care personnel available in the Trust. However, in a field such as HD where understanding of the condition is limited, sharing information about treatments and therapies is essential to providing the highest quality care. The third sector already seems to be playing a key role in the provision of care and support yet there is limited understanding of their contribution, status or how they fit with services provided by the NHS.

This study has provided some evidence about how people manage their diagnosis and new knowledge of HD. However further research is needed to gain a better insight into how people subsequently live with that new knowledge, how, when and if that genetic information is used and shared. The secrecy and silence surrounding the knowledge of HD in families has been touched on in this thesis and has been shown to be an issue for many families. Better understanding of these issues may contribute to a shift in attitudes towards HD.

Further research is also needed into the ways in which people manage their own condition and how this might work in the longer term alongside easy access to service provision. There is very limited understanding of how people with HD wish to be given information about their condition and managing independently. The efficacy of self-management programmes for family carers should also be explored. In addition, the issues for family carers warrant further

examination. This should address the effects of turning homes into spaces for providing care, and the impacts of a move to residential care for the person with HD.

WHAT THIS STUDY ADDS

This study has contributed to the field of HD in two key ways. First, this is the first longitudinal qualitative study which has included multiple methods of data collection and multiple perspectives to gain a picture of living with HD. This study has shown this to be an appropriate and successful methodological approach within the field of HD. Second, is the contribution to the knowledge around care provision and the needs of those with HD, their families and the challenges for the staff delivering care.

Due to their diminishing abilities patients are often excluded from research in HD. This study has undertaken specific adaptations to be flexible enough to include not only patients, but also their families and staff members. The study has focused on context and depth to create multiple detailed case studies and consequently each case has contributed to a wider understanding of living with HD.

Findings have highlighted the importance of the multidisciplinary team for those with HD. Examples of this working well across different sites and in different formats have been provided alongside some of the challenges for the health and social care professionals involved. Quality care was shown to be holistic, incorporating the needs of the whole family. This was facilitated by a health or social care professional acting as a single point of contact, or key worker. This person got to know the patient and their family, building trust and an overarching view of the many components that individual family faced while living with HD. The need for specialist knowledge of HD was illustrated as vital to supporting patients and families.

Long-term caring means that even when a patient moves to a residential care home the burden for family carers is not necessarily lifted but is merely shifted. This was shown to leave family carers in a liminal state whilst they re-balanced

their lives as a single person and shifted from an active 'carer' role to a more passive 'caring' role. In line with the issues around choice and decision making some patients did not always want to engage with services for management. Whilst care planning was desirable it was not always possible and care that was flexible and responsive was needed to attend to crisis situations.

This study has highlighted that HD poses unique issues for service provision and care planning due to the complex nature of the disease and the rarity of the condition. It remains unclear what services are currently being provided across the UK and how well these services work in terms of providing quality care to families affected by HD. This study has demonstrated three different types of care provision, each generally working well. These three services highlighted the role of the clinical nurse specialist for HD as a co-ordinator of care and the wider multidisciplinary team; the essential input from the third sector in the provision of the RCA to support a consultant and provide community out-reach; and the importance of experienced care in the long-term residential setting. All these types of services illustrated key workers, HD specific knowledge and continuity of care as essential to quality care for those trying to balance the delicate equilibrium of living with HD.

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APPENDICES

PUBLICATIONS FROM THIS THESIS

Case study example for Huntington's disease in: Woodward S and Mestekcy A (2011) *Neuroscience Nursing – evidence-based practice*. Oxford: Wiley Blackwell p452, case example from Elizabeth and James.

Wilson, E, Pollock, K and Aubeeluck, A (2010) Gaining and maintaining consent when capacity can be an issue – a research study with people with Huntington's disease. *Clinical Ethics* Vol5(3):142-147 Cited on the NIHR website as 'Valid informed consent resources' http://www.crncc.nihr.ac.uk/training/courses/other/vic_resources

Wilson, E (2009) 'Patients and families' in Byrne J, McNamara P, Seymour J, and McClinton, P (eds) *Palliative Care in Neurological Disease: A team approach*, Radcliffe, Oxford pp85-98, case example from Sarah and Max

Wilson, E, Elkan, R, Seymour, J and Almack, K (2008) A UK literature review of progressive long-term neurological conditions. *British Journal of Community Nursing* vol13(5):206-212

Aubeeluck A, and **Wilson E**, (2008) Huntington's disease. Part 1: essential background and management. *British Journal of Nursing* Vol17(3): 146-151

Service users



Information Sheet

Exploring the palliative care needs of service users with Huntington's disease

My name is **Elli Wilson**

I am a researcher at the **University of Nottingham**

I would like to **invite you** to take part in a **research study**

Service users

The study aims to **find out** what it is **like to live with Huntington's disease**

Please take the time to read this information booklet

Please **ask any questions** you want before you decide

Discuss it with others if you wish.

What is the purpose of the study?

To try and **understand** what it is **like to live** with **Huntington's disease**

By finding out what it is like it can **help us** to **inform staff** how to give the **best possible care and services** to help people with Huntington's disease to live well.

The study will focus on **your thoughts and opinions** about your **current care** and **your expectations** for your care in the future.

Why have I been chosen?

You have been chosen because you have **Huntington's disease** and have been **accessing these services.**

Do I have to take part?

No. Taking part is **voluntary**

What will the study involve?

- **Spending some time** with you **during your daily activities**
- **Talking to you** and writing down some **notes**
- Interviewing you in a private room
- **Recording** what you say
- **Talking** to your **relatives**

This will be for a **few hours** at a time

Service users

What are the disadvantages of taking part?

You may **not want me to be there**

You may **not want to talk to me**

HOWEVER:

I will always **ask you first**

You can **stop** at any time

You can **start** again **any time**

Talking about your condition **may upset you.**

If this happens, I will make sure there is **someone to support you.**

What are the possible benefits of taking part?

The study may **help us understand** more about living with **Huntington's disease**

The study may **help to improve services** provided to people with Huntington's disease and their families in future.

Will my taking part in this study be kept confidential?

Yes. Anything I see or hear will be **kept confidential**

I will **not share** it with anyone else

Your name will not be used in my notes or on the recordings.

Service users

What will happen to the results of the research study?

The findings will be written up **in a PhD**

Your name will be kept secret

You will be given a summary **to keep**

Who is organising and funding the research?

Elli Wilson **PhD Student**

The Sue Ryder Care Studies Centre at the University of Nottingham.

Who has reviewed the study?

This study has been reviewed and approved by the **Nottingham Local Research Ethics Committee.**

Where can I get further information?

You can ask me – **Elli** Wilson

I can be contacted on **0115 82 31201**
eleonor.wilson@nottingham.ac.uk

Thank-you

service users



Consent Form

**Exploring the palliative care needs of service users
with Huntington's disease.**

service users

- I have seen the **information booklet** about this research study, dated 16/05/07 (version 4)

YES



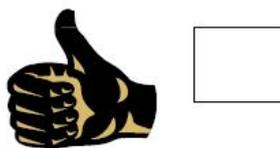
NO



- **I have talked** with Elli Wilson about the study



YES



NO



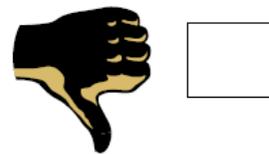
service users

- We have **looked at the information** together

YES

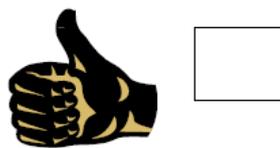


NO



- **My questions have been answered**

YES



NO



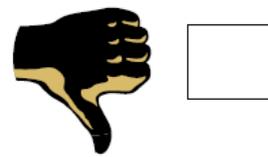
service users

- I **understand** what is involved in the study and my part in it

YES



NO

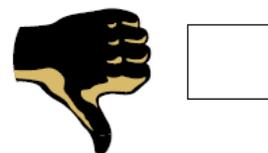


- I **understand** that if I choose to take part, it is **my free choice**

YES



NO

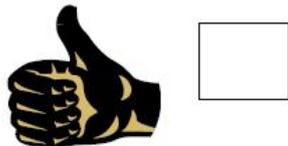


service users

- I **understand** that **I can stop** at any time and my care and treatment will not be affected



YES



NO



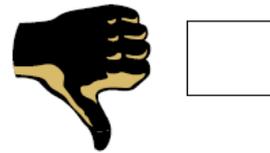
service users

- I **agree** to take part in the study

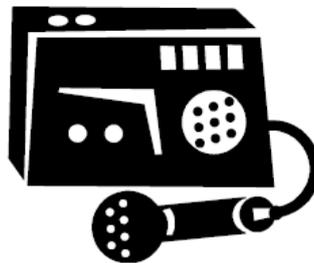
YES



NO



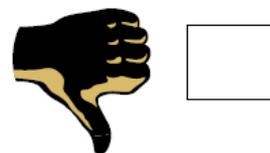
- I **agree** to be audio **recorded**



YES



NO



service users

- I **agree** that the information may be used in **Elli's PhD** and published **reports**



- I will **not** be named or identified

YES



NO



service users



Name of person agreeing to take part:

Signature: _____

Date: _____

Name of person taking/witnessing consent:

Signature: _____

Date: _____

APPENDIX D: RELATIVES INFORMATION SHEET

Informal carers



Information Sheet for Informal Carers taking part in case studies

**Exploring the care needs of people affected
by Huntington's disease.**

You are being invited to take part in a research study. Before you decide whether to take part it is important for you to understand why the research is being done and what it will involve. Please take time to read the following information carefully and discuss it with friends and relatives if you wish. Ask me if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish to take part.

What is the purpose of the study?

The study aims to gain an understanding of what it is like to live with someone with Huntington's disease both at home and in a specialist care centre. Asking informal carers about their own needs and those of the person they look after will help us to inform staff where they are providing good care and where they may need to raise standards.

Why have I been chosen?

You have been chosen because you have a relative who has been diagnosed with Huntington's disease and has lived this condition for a number of years.

Do I have to take part?

It is up to you to decide whether or not to take part. If you do decide to take part you will be asked for your written consent at the start of the study. However, you are still free to withdraw at any time and without giving any reasons. Your relative's medical and nursing care will not be affected by whether or not you decide to take part in the study.

What does the study involve?

In order to talk about your views and opinions of your relative's care you will be asked to take part in a series of interviews. As I want to gain as much insight as possible about living with Huntington's disease I may also ask to observe some parts of your relative's care such as a physiotherapy session, a consultation or daily activity. During this time I will spend some time with you and your relative talking to you and taking some notes about what is happening. However observations will only take place when both you and your relative are fully aware that we are doing. I will also ask permission from anyone else who is there during this time, for example; the speech therapist. During this time you may ask me to stop or leave the room with out giving any reason. Before starting the each interview I will ask your permission to audio record to help me remember what you have said accurately. Your name will not be on the recording or any written reports. Interviews might take up to an hour of your time, however periods of observation may vary. Interviews can take place in your home or another place of your choosing at a time suitable to you.

What are the possible disadvantages and risks of taking part?

The experience of illness is often a cause of anxiety for all people involved. There is a possibility that the interviews will touch on upsetting topics. However, you will not be under any pressure to answer questions or talk about topics that you prefer not to discuss. The interviews will focus on your thoughts and opinions about the care currently being provided and your expectations of future care for your relative.

What are the possible benefits of taking part?

We hope that the results of the study will contribute to an improvement in the services provided to users and their relatives in future.

Will my taking part in this study be kept confidential?

All information which is collected about you and your relative during the course of the research will be kept on a password protected database and is strictly confidential. Any information about you which leaves the research unit will have any identifiable details removed so that you cannot be recognised from it.

What will happen to the results of the research study?

The information gained in this study will be used to help health care staff provide good quality care for people with long-term neurological conditions and their families. The results of the research will be made more widely available through journal publications and conference presentations. You will not be identified in any report or publication resulting from the research.

Informal carers

Who is organising and funding the research?

Elli Wilson – PhD student
Division of Nursing at the University of Nottingham

Who has reviewed the study?

This study has been reviewed and approved by the Nottingham Local Research Ethics Committee.

Contact for Further Information

Elli Wilson

B63b, Division of Nursing
University of Nottingham
Queens Medical Centre
Nottingham
NG7 2HA
Tel: 0115 8231201
E-mail: eleonor.wilson@nottingham.ac.uk

APPENDIX E: RELATIVES CONSENT FORM

Informal carers

Identification Number:



CONSENT FORM

INFORMAL CARERS

Exploring the palliative care needs of service users with Huntington's disease

Please initial box

1. I confirm that I have read and understand the information sheet dated 16/05/07 version 3 for the above study and have had the opportunity to ask questions.	
2. I understand that my participation is voluntary and that I am free to withdraw at any time, without giving any reason, without mine or my relatives' health care or legal rights being affected.	
3. I agree to the interview being recorded for the purposes of the research. I understand that I may ask for the recording to be stopped at any time.	
4. I agree to the use of extracts from the interview being used in published reports and presentations resulting from the research. I understand that all personal details will be removed and I will not be identified in any published work.	
5. I agree to be observed during appropriate times and when my permission has been sought.	
6. I agree to notes being taken during these observations	
7. I confirm that I am aware that I can asked for observations to be stopped at any time without giving any reason.	
8. I agree to take part in the above study	

Name

Date

Signature

Researcher

Date

Signature

version 3 17/05/07

APPENDIX F: HEALTH AND SOCIAL CARE PROFESSIONALS INFORMATION SHEET

Health and social care professionals



Staff Information Sheet

**Exploring the palliative care needs of service users
with Huntington's disease.**

Introduction:

My name is Elli Wilson, I am a PhD student at the University of Nottingham. This work aims to explore the palliative care needs of people with Huntington's disease.

You are being invited to take part in this research study. Before you decide it is important for you to understand why the research is being done and what it will involve. Please take time to read the following information carefully and discuss it with others if you wish. Please ask me if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish to take part.

Thank you for reading this.

What is the purpose of the study?

The study aims to gain an understanding of what it is like to live with someone with Huntington's disease both at home and in a specialist care centre. The study aims to include the views and experiences of a range of key health and social care professionals providing care to people with Huntington's disease. I will also be gaining views from service users and their informal carers about their experiences of care and support.

Why have I been chosen?

You have been asked to take part because you have been identified by someone with Huntington's disease as a key person involved in their care.

Do I have to take part?

You do not have to take part in this study and if you chose to do so it is entirely voluntary so it is up to you to decide whether or not to take part. If you do decide to take part you will be given this information sheet to keep and be asked to sign a consent form. If you decide to take part you are still free to withdraw at any time and without giving a reason. A decision to withdraw at any time, or a decision not to take part, will not affect you or your work in any way.

What will happen to me if I take part?

If you do decide to take part in this study you will be asked to take part in an interview. This may take up to an hour of your time. The discussion will be recorded and can be held in a place of your preference. I want to talk to you about your views and your opinions of providing care to someone with Huntington's disease so there is no need for you to do anything to prepare.

Health and social care professionals

Will my taking part in this study be kept confidential?

Any information or opinions you give during the discussion will be anonymised. Recordings will only be listened to by the researcher and will be kept in a locked office at the University in Nottingham. You will be assigned a code so that your name is not used at any time. Any distinguishing information will be edited from any written comments so that you cannot be identified in any way.

What will happen to the results of the research study?

The results will be used to help inform providers of services about the challenges facing health care professionals delivering care to people with Huntington's disease, and help them acknowledge good practice and improve care.

Contact for Further Information

If you have any questions please do not hesitate to ask. The contact number for further information is
0115 8231201

Eleanor Wilson

APPENDIX G: HEALTH AND SOCIAL CARE PROFESSIONALS CONSENT FORM

Health and social care professionals

Identification Number:



The University of
Nottingham

STAFF CONSENT FORM

Exploring the palliative care needs of service users with Huntington's disease

Please initial box

1. I confirm that I have read and understand the information sheet dated 16/05/07 version 1 for the above study and have had the opportunity to ask questions.	
2. I understand that my participation is voluntary and that I am free to withdraw at any time, without giving any reason, without my work or legal rights being affected.	
3. I agree to the interview being recorded for the purposes of the research. I understand that I may ask for the recording to be stopped at any time.	
4. I agree to the use of extracts from the interview being used in published reports and presentations resulting from the research. I understand that all personal details will be removed and I will not be identified in any published work.	
5. I agree to take part in the above study	

Name of interviewee

Date

Signature

Researcher

Date

Signature
