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UNIVERSITY OF SOUTHAMPTON

FACULTY OF HEALTH SCIENCES

**Exploring the impact of a diagnosis of Huntington's Disease on couple
relationships**

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Thesis for the degree of Master of Philosophy.

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ABSTRACT

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Exploring the impact of a diagnosis of Huntington's Disease on couple relationships

Christopher Vincent

Abstract-This study explores the impact of a diagnosis of Huntingtons Disease (HD) on couple relationships. A qualitative research method was used, based on the Free Association, Narrative and Interview Method, in order to interview three couples and four individuals. The couple relationships examined were at different stages of the HD illness trajectory, ranging from one couple where a diagnosis was made eighteen months before interview to two couples whose partners had died following several years of illness. From a thematic analysis of interview content, three over-arching themes and ten sub-themes were identified. The first over-arching theme refers to the ways that couples first became fully aware of the family history of HD and the implications this knowledge had for their lives and others within their families. The second overarching theme tracks the different ways that this knowledge was handled by the couple themselves and with other family members. The third overarching theme identifies key dilemmas faced by couples in managing the transition from a pre-illness to an illness dominated relationship. These dilemmas were-a) balancing anxious concern with a respect for independence, b) managing the loss of a sexual relationship and c) managing the move into residential care. All three of these challenges can be understood as being linked to how couples regulate emotional and physical distance during the course of the illness trajectory.

Key words:-Huntington's disease, couple relationships, family history, communication, distance regulation, residential care, qualitative research.

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Biography of Author.

I am a couple psychotherapist registered with The British Psychoanalytic Council and have worked exclusively in private practice since 2006. I hold a portfolio of work related commitments particularly those connected with training and supervision. I am Consultant Visiting Lecturer at Tavistock Relationships, London and I contribute teaching and supervision to organisations in this country and abroad. In addition to my specialist interest in couple therapy and family related matters, I completed a training, in 1994, with the British Association of Psychotherapists to work intensively with individuals.

I completed a degree in PPE at Oxford University in 1967 and, after undertaking a period as a Community Service Volunteers, I qualified as a social worker in 1971 from Exeter University. Following spells in community and paediatric social work, I joined what is now Tavistock Relationships in 1977 where I was a senior member of staff and Research and Publications Coordinator until 2002. On leaving TR, I worked until 2006 as a part time Consultant Psychotherapist in an NHS hospital based psychiatric service.

Declaration of Authorship

I, CHRISTOPER VINCENT]

declare that this thesis and the work presented in it are my own and has been generated by me as the result of my own original research.

‘Exploring the impact of a diagnosis of Huntington’s Disease on couple relationships.’

I confirm that:

1. This work was done wholly or mainly while in candidature for a research degree at this University;
2. Where any part of this thesis has previously been submitted for a degree or any other qualification at this University or any other institution, this has been clearly stated;
3. Where I have consulted the published work of others, this is always clearly attributed;
4. Where I have quoted from the work of others, the source is always given. With the exception of such quotations, this thesis is entirely my own work;
5. I have acknowledged all main sources of help;
6. Where the thesis is based on work done by myself jointly with others, I have made clear exactly what was done by others and what I have contributed myself;
7. None of this work has been published before submission.

Signed:

Date:

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Sharing experiences with an unknown researcher requires great trust in the process. I have been acutely conscious of the trust vested in me by the anonymous participants in this study. I hope that my respect for and gratitude to them is reflected in the way that I have recounted their experiences of living with HD.

Definitions and Abbreviations

Definitions/Abbreviations: The following abbreviations will be used for convenience. Huntington's disease will be referred to as HD and the charitable organisation representing the interests of HD families, the Huntington's Disease Association, as the HDA. The terms 'patient' or 'client' will only be used where this reflects contexts where these terms have been employed, for e.g. in medical or counselling journals.

Chapter 1. Introduction

1.1 Background to the study

Investigating the connections between Huntingdon's disease and couple relationships is a specific research focus which grew out of a more general interest in the psychology of later years and the role that dementia plays in couple and family life. Several elements came together to encourage initiating a research study in this area.

The first was a clinical interest arising from seeing increasing numbers of middle and older aged clients in my couple psychotherapy practice, some of whom had, or were concerned to have, neurological problems and aware that my theoretical understanding of these phases of life and conditions was not as developed as it needed to be. Psychoanalytic theory, which has formed the backbone of my approach to therapy, is rich in describing the developmental phases of childhood and early adulthood but is only slowly building a body of theory to match the experience of an ageing adult population some of whom turn to therapists for help.

Psychoanalytic theory lays emphasis on how the past shapes current experience and underestimates how the anticipation of future frailty and eventual mortality influences the present. There are some notable exceptions to this general statement. Jung held the view that mid life and old age offered important development opportunities and his ideas stood in marked contrast to Freud's pessimism about the potential of mid and later years being creative; for this reason, Freud once claimed that psychoanalysis should not be offered to older applicants (Freud 1905). Jung thought that mid-life heralded a change when an individual's preoccupations turned from a focus on establishing an identity and place in society to becoming more inwardly reflective. He thought that this shift was prompted by an awareness of personal mortality and the existential questions about a place within the cosmos which then follow (Jung 1972). The connections between the anticipation of mortality and mid-life were pursued by Jaques (Jaques 1965) writing from a Kleinian perspective. He argued that if individuals are to come to terms with their own mortality they have to celebrate what they have achieved in life and mourn what has not been possible. This reckoning with the past, where positives and negatives are weighed in the balance, marks a shift from the paranoid-schizoid position (where the world is seen as either being all positive or all

negative) to the more nuanced depressive position where mixed feelings can be held onto simultaneously. In addition, the psychoanalytic community has benefited from edited collections from this developing field (Junkers 2006; Davenhill 2007) and from the work of psychogerontologists (Kitwood 1997; Coleman and O'Hanlon 2004).

Psychoanalytic theory in the Object Relations tradition is essentially a set of theories about mind and, in my view, underestimates the significance of the body in its theories. This lack becomes apparent when working with older people for whom bodily changes, whether arising from ill health or progressive physical weakening, can assume great significance in mental life. 'Dialogues' with the body can be as significant as dialogues with real people and/or dialogues with figures from the past held in the imagination. In the specialist field of couple psychotherapy, there is an absence of clinical theory that locates bodily preoccupations resulting from ill health within the array of other 'objects' that can complicate couple functioning. The experience of a failing body constituting an intrusive and distracting third presence in a two person interaction arises in work with some older clients and seeking to understand this and other related phenomena has impelled me to want to understand more about the relationship between illness, ageing and couple functioning.

Therapeutic practice within the broad psychodynamic field views the relationship between client and therapist as central to the work. The interactions between client and therapist can both illuminate the nature of the client's problems (client problems get re-enacted in the consulting room) and can be the means by which problems are transformed or are managed in new ways. Either way the therapist gets caught up emotionally and cognitively with his client's difficulties and utilizes his emotional resonance to help understand client experience. This idea is counter to Freud's notion of the psychoanalyst acting as a 'blank screen' and, instead, asserts that, through processes of projection and identification, a therapist can use his own emotional reactions as evidence to complement other sources of data in making clinical judgments. The difficulty in working in this way is that the therapist's personal experiences can both help or hinder. They can help if they assist with making a helpful identification with another's experience. They can hinder if they are imposed on another's experience in a way that denies that experience. Ensuring that the former rather than the latter prevails requires that the therapist knows him or herself as well as possible particularly with regard to those aspects of personal experience that are close to those of the client group.

Along with most therapists and counselors, I trained as a relatively young person. None of this training equipped me for facing the challenges of becoming old (I am now 70) or knowing what it would be like to become seriously ill as I was during 2012. I am, therefore, conscious that one of the drivers for gaining an interest in illness and ageing is the personal benefit this knowledge affords me which hopefully I am able to put to some use in my clinical work. As a researcher, I am ‘wounded’ in the sense that Frank (Frank 2013) writes about.

A second factor which encouraged me to think about a research project was the possibility of drawing on research results to develop training materials and publications for professional and service user audiences. In 2009 the Government launched the National Dementia Strategy, updated later that year by the Implementation Plan (Department of Health 2009). This plan emphasised the need for the provision of good quality early diagnosis and intervention for all patients and their families and, as part of achieving this aim, the Implementation Plan recognised the need for an informed and effective workforce. I believed that knowledge about the impact of diagnoses on the relationships of patients and their partners might contribute towards an informed and effective workforce.

A third factor which influenced the decision to develop a research project as part of a psycho-social training programme was a longstanding interest in wanting to be clear about the research status of small scale, in depth, qualitative studies. Gaining some clarity about this subject seemed to me to be important when any results from the sort of small scale study being envisaged would be challenged by professionals and others whose research sympathies were likely to be with larger scale quantitative paradigms.

My subject interests and research interests came together in 2008 when I was introduced to a small NHS psychiatric team which offered diagnostic and support services to families where one adult had an early onset dementia. These conditions included Fronto-temporal dementia, Lewy Body disease, Vascular dementia, Alzheimers disease, Alcohol related dementia and Huntington’s disease (Thompson and Coates 2006). I was encouraged from these meetings to consider developing a research study within the NHS looking at the impacts of early onset dementia on couple functioning. As a result of a series of discussions with NHS and voluntary sector organisations, it became apparent that the institutional and technical support

needed for the study would best be provided through the Faculty of Health Sciences at Southampton University where I enrolled as a part time research student in 2009.

The choice of a study focusing on Huntington's disease was made in the light of findings from a Scoping Review which considered the possibility of a more broadly based study looking at an array of early onset dementias and their impacts on couple functioning. The reasons for narrowing the focus on HD and couple relationships are described later but because of the unique character of Huntington's disease and its relative unfamiliarity to lay and professional persons, an early introduction to the condition is now given.

1.2 Huntington's disease, St Vitus's dance, 'the migrims' and 'the magrums'.

Like many illnesses, HD arrives unbidden into the lives of those it affects. "Illness is the night-side of life, a more onerous citizenship. Everyone who is born holds dual citizenship, in the kingdom of the well and in the kingdom of the sick. Although we all prefer to use only the good passport, sooner or later each of us is obliged, at least for a spell, to identify ourselves as citizens of that other place", Sontag (1979 p3).

For some, a family history of the disease provides forewarning of its emergence while for others it comes as a shock. Whether there is forewarning or not, because of the distinctive ways it affects patients and their offspring, the disease initiates a devastating disruption to pre-existing ways of living. An account of the disease from a conventional medical perspective will give some idea of the nature and scale of the unique challenges HD poses for the mental wellbeing of individuals, couples and families and the demands it makes to reside in this 'other place'.

Huntington's disease is named after a family doctor, George Huntington who, like his father and grandfather before him, practiced on Long Island, U.S.A. in the early and mid-19th century. He had grown up an acute observer of the patients served by his father and grandfather, having travelled around with them on their domiciliary visits (Wexler 2008). Later, in his own practice, he drew on his acute observational skills to describe an inherited pattern of neurological and psychiatric symptoms which he published in a paper, On Chorea,

in 1872 (Huntington 1872). In this paper he described chorea as “the dancing propensities of those...affected” in whom there “seems to be some hidden power, something that plays tricks, as it were, upon the will” (Huntington 1872 p109) .While much of the paper is focused on the treatment of chorea in general, he devotes the final page and a half to describing the particular form of neurological disease that bears his name. He identifies three characteristics of the disease. The first is that the disease is inherited and is passed from generation to generation; “it is spoken of by those is whose veins the seeds of the disease are known to exist, with a kind of horror.” (Huntington 1872 p111). In recognising the inherited nature of the condition he also observed that some children became carriers of the disease while others did not and he noted that for those children who were not affected the line of inheritance was broken. The second characteristic he identified was the tendency for the condition to lead to insanity and suicide-“as the disease progresses the mind becomes more or less impaired, while in others, mind and body both gradually fail until death relieves them of their sufferings” (Huntington, 1872 p112). Finally, he thought that it was a disease of middle adult life appearing after the age of forty and was incurable.

The main features of the disease described by Huntington are broadly accurate in the light of what is known today. HD is a condition that, within the medical world, bridges the gap between psychiatry and neurology. Paulson and Albin (2011) comment that physicians are continually astonished by the diverse array of symptoms and signs that the affected person develops so that each person with HD is clinically unique. It is a disorder typically, though not exclusively, emerging in middle age with most people developing the condition between the ages of 35 and 55 years (Quarrell 2008). As an autosomal dominant genetic disorder the children of any affected individual have a 50% chance of inheriting the disease. The age of onset has a normal distribution across the lifespan with the onset in any particular individual being largely determined by the nature of their gene mutation (Paulson and Albin 2011). This particular neurological feature emerges from the gene test that has been available since 1993 and is offered after counselling. Tested individuals can be given what is known as their CAG count although clinical judgment varies as to whether patients should be given their specific count or a general indication as to whether it is abnormal or normal (Quarrell 2008). A CAG count below 27 is unequivocally normal; a result between 27 and 35 is normal but may increase in the next generation of offspring. A result between 36 and 39 is abnormal but the disease may not emerge in a patient’s lifetime. Above 40 the result is unequivocally abnormal

and a person with this score who has one parent with the disease will develop the condition but it is not possible to state when this will start (Quarrell 2008).

While the presentation of the disease varies widely, Paulson and Albin describe the classic clinical triad as being 1) a progressive movement disorder, most commonly chorea: 2) progressive cognitive deterioration culminating in dementia and 3) various behavioural disturbances often starting before diagnosis (Paulson and Albin 2011). It was in recognition of all three of these components that in modern times the condition has come to be called Huntington's disease rather than Huntington's chorea, the term originally emphasised in the 1872 paper. It is possible to locate all three of these elements in the 15-20 year lifecycle of the disease's progression.

How and when the disease emerges is described differently in different studies. There is invariably a lead time before the condition is diagnosed and in order to ascertain features of this pre-diagnosis stage Kirkwood et al. (2001) asked the 1st degree relatives of one thousand, two hundred and thirty eight individuals with a HD history of at least six years duration to complete a questionnaire recording their recall of the earliest symptoms. Kirkwood found that the earliest symptoms were involuntary physical movements, followed, at the second stage, by the emergence of mental and emotional symptoms. A third stage was identified in which clumsiness, sexual problems, lack of motivation and paranoia appeared. Then a variety of additional problems were recorded including unsteadiness and difficulty walking, a problem in holding onto objects, sleeping irregularity, hallucinations, intellectual decline and memory loss. While Kirkwood's sample identified involuntary physical movements as one of the first signs, Martinez-Horta et al. (2016) and van Duijn et al. (2008) observed the emergence of psychiatric symptoms, especially apathy, in the stage before there are problems with physical movements. These studies resonate with the findings of a study by Halpin (2011) who discusses the frequent misdiagnosis of Huntington's disease in the pre-diagnostic or prodromal stage. He suggests that patients are frequently assessed by mainstream psychiatrists who ascribe their patients a psychiatric diagnosis. The suggestion in Halpin's paper is that the pathways to care in the health services help define the nature of early symptoms. In his study many respondents reported their relatives having been 'locked away' and given inappropriate treatment as a result of misdiagnosis.

In summary, the early stages of the disease may involve:-

- Jerky chorea movements, a slowing down of voluntary movements, slurring of speech (Quarrell 2008)
- Involuntary physical movements, mood changes, clumsiness, sexual problems, apathy and paranoia, sleeping irregularity, cognitive impairments (Kirkwood 2001)
- Neuro-psychiatric symptoms at early stages (Martinez Horta 2016; van Duijn 2008).
- Slaughter et al. (2001) in a review of several studies concludes that depression has a general prevalence rate of 30% for all HD patients but Epping and Paulsen (2011) conclude that depression has the highest prevalence rate in the early stages when symptoms become manifest and will also emerge in twelve months of the prodromal stage before motor symptoms appear. Codori et al. (2004) confirm this picture finding that depression runs at a rate of 20% in the year after getting a positive test which compares to a rate of 12.6% for those with a negative result. This study concludes that a positive predictive test result is not psychologically benign and the authors recommend that those tested positive should have a psychiatric follow up in the year following testing as a matter of routine. This view is repeated by Andersson et al. (2016) in the conclusion of a case study mentioned later in the Literature Review.
- Suicide is a significant risk for the HD population as a whole. Bindler et al. (2010) assess it to be four times higher than in the general population. Epping and Paulsen (2011) state that within the HD population it runs at a rate between 5-10%. Paulsen et al. (2005) have identified two critical periods when the risk of suicide peaks. These are immediately after a formal diagnosis is made and at a later stage when independence markedly deteriorates. Other factors associated with the risk of suicide include unemployment and a psychiatric history in the 5 year period before diagnosis (Almqvist et al. 1999), having no offspring and being single or divorced (Lipe et al. 1993) and misusing alcohol and/or drugs (Wetzel et al. 2011).

Quarrell in his brief outline of the disease's progression suggests, somewhat guardedly, that it is possible to think of middle and final stages. In the middle stages he describes the physical, cognitive and emotional symptoms worsening. Physically, chorea movements are likely to be more pronounced and there may be difficulties in maintaining balance. Speech may become more slurred and swallowing may prove difficult. In terms of cognitive deficits, concentration levels may fall away although long term memory tends to remain good. Social, emotional and behavioural problems become more pronounced. These may include depression, apathy,

irritability, aggressive outbursts, heightened anxiety and impulsive behaviour including sexual disinhibition. In the later stages of the illness there may be a tailing off of the chorea and, instead, the body may be prone to stiffness and rigidity (dystonia). General mobility becomes restricted and residential care will become necessary as the patient becomes unable to undertake basic bodily tasks including washing, toileting and eating.

1.3 HD and society: how HD has been viewed over time.

It will be apparent from a description of the medical symptoms that there will be a complex relationship between the biological realities of HD and the social contexts in which it appears. Wexler states this clearly: “How disease is imagined, and how sufferers are portrayed, helps shape the experience of illness” (Wexler 2008, p125). There is now an evolving literature on how HD has been understood within different social groupings and at different historical periods. Wexler suggest that while physicians did not see HD as a single disease until the mid-19th century, there is evidence to suggest that non-medical people had long recognised the condition as an illness. It was described variously; in some places it was described as St Vitus’s dance connecting St Vitus, the patron saint of dance, with the chorea or dance like involuntary movements. In other places it would have been referred to in more colloquial terms. Wexler points out that in parts of New England and New York it was called the “migrims”, a term similar to an old dialect word used in Lancashire, the “magrums”, meaning “in a state of temper or rage” (Wexler 2008 p XIX). Sometimes the name attributed to the disease reflected the localities where the disease was concentrated and this connection was mentioned in one of the research interviews where the interviewee referred to “X disease”, where “X” referred to a small village in the South East of the UK where a number of HD families resided.

Huntington’s seminal paper in 1872 is seen as the point in history when the disease came to be understood by the medical community in relation to its biological origins. The identification of the disease coincided with a broad scientific debate at that time which built on Mendel’s pioneering discoveries about genetic inheritance. One outcome of this debate was the claim made by Francis Galton in the late 19th century that education and the environment could not influence biological heredity and that improvement of the human race depended on encouraging the ‘fittest’ and discouraging the “unfit” to procreate. So the

pseudo-science of eugenics was born, the echoes of which stretched long into the 20th century. The combination of ignorance about the genetic basis of HD (the identification of the malfunctioning gene was not identified until 1983) and public misunderstanding about the nature of some HD symptoms which can be disconcerting to the onlooker, created a knowledge void filled by the stigmatisation of HD patients and their families. Two papers are cited as particularly influential in this process. Vessie (1932) argued that bad or amoral personality traits (including the suggestion that women might be witches and that men might be criminals) were inherited down the generations and he purportedly traced these connections back to the first immigrants from the UK to New England in the 17th century. This theme and research method was repeated by the British neurologist, Critchley (1934) who suggested that the East Anglian ancestors of the New England HD population were similarly flawed individuals. Subsequent research has refuted the veracity of Vessie and Critchley's findings (Wexler 2008) and yet there remain remnants of this stigmatisation into recent times.

Wexler summarises some of the evidence given to the Commission for the Control of Huntington's Disease and Its Consequences (1976-77) set up with the backing of the U.S Congress. One striking conclusion from the testimony given to the Commission was that from the 1940s to the mid-1970s, young people at risk from HD received explicit warnings from physicians and counsellors not to have children; "some physicians were offering direct eugenic advice to their patients well into the 1970s" (Wexler 2008 p.178). As late as the 1980s Critchley continued to maintain that people with HD showed reprehensible behaviour including "intermarrying, illegitimacy and incest" (Critchley 1984 p.725). He cited the high costs of each patient and lamented that HD was not a notifiable disease as he thought its prevalence could only increase, which it has done but not for the reasons he predicted. In more recent times, Williams et al. (2010) reported on research to evaluate stigma and discrimination experienced by 'at risk' individuals who had been tested for HD and had either received a positive or negative result. Four hundred and twelve of the four hundred and thirty three participants drawn from Canada, the U.S and Australia provided narrative accounts of their experiences after testing. The overarching theme of Information Control was identified by the researchers to capture the participants' diverse experiences. These included a loss of control of private medical information when professionals, friends or relatives passed on information without consent and this information was then used against the patients' interests in insurance and employment situations. Faced by this discrimination some participants

reported fighting back and becoming more open about their medical histories while others reported becoming more guarded and wary about making medical disclosures. It is then not surprising that some affected individuals who adopt an avoidant strategy turn against social engagement, feel diminished self-confidence and seek to hide knowledge of HD from themselves and from others (Wexler 2010).

The identification of the problematic gene in 1983 and, later, the introduction in 1993 of a test to identify those individuals who are likely to develop the disease has, according to Huniche (2011), generated a moral consensus or culture encouraging at risk young adults to engage with genetic information, services and technologies. She argues that the culture places pressure to confront procreational risks before trying for children and that to do so expresses greater moral responsibility than avoidance.

1.4 How prevalent is HD and who bears the costs?

In 2010 an All Parliamentary group for HD was launched and, to help in their work, a call was put out to establish the scale of the illness within the UK (Spinney 2010). Two studies have provided clarification. Evans et al. (2013) collected data on all records of patients aged over twenty one years with a diagnosis of HD from the UK GP Research data base. The results showed that prevalence increased from 5.4 per 100,000 of the population in 1990 to 12.3 in 2010. Geographically the London region showed the lowest rates at 5.4 while the highest rates were in the North East (18.3) and Scotland (15.8). The study concluded that there were 5,700 patients with HD in 2010 which represented a doubling of the recorded HD population in medical records between 1990 and then. This increase was most notable in older patients who are surviving longer, partly as a consequence of improved care but also reflecting the general increase in life expectation of the general population. Morrison (2010) undertook a review in Northern Ireland and recorded a similar increase in prevalence rates from 6.4 to 10.6 per 100,000 between 1991 and 2001. Internationally, there is wide variation in prevalence rates. Highest rates are recorded in Western Europe, North America and Australia while the lowest rates are found in Asia (Rawlins et al. 2016). These figures are of diagnosed cases and do not take account of the large number of individuals who have not developed symptoms nor of those who are symptomatic but have, for various reasons, chosen not to seek medical care. Rawlins is quoted as saying that “The rule of thumb is that the

number of pre-symptomatic people in a population is roughly twice the number of symptomatic” (quoted in Spinney, 2010).

While the prevalence of HD in the community since 1990 has increased, Rawlins et al. (2016) argue that this has not been the result of an increased incidence of HD but is likely to be the result of a) better knowledge among doctors to diagnose correctly, helped by genetic testing b) a reduction in patients feeling too ashamed to approach medical services and c) patients surviving longer, partly as a result of better medical and community care but also as a result of extended longevity.

HD is one of the early onset dementias and the burden of care placed on all families caring for a person with dementia has been evaluated by health economists. Luengo-Fernandez et al. (2010) estimated that 55% of the total costs of caring for dementia patients falls on unpaid carers, 40% are social care costs and just 5% are health care costs. Jones et al. (2016) has figures showing a greater proportion of costs falling on families with HD. He estimated, on the basis of examining the social and medical costs of one hundred and thirty one individuals drawn from the European Huntington’s Disease Registry Study, that 65% of the costs are covered by informal carers at all stages of the disease. He and colleagues conclude that their analysis backs the case for improving community based support services for HD families. This broad picture of families bearing the major proportion of care costs is compounded by the effects of HD on family incomes. McCabe et al. (2008) investigated the impact of Multiple Sclerosis (MS), Motor Neurone Disease (MND), Parkinson’s disease and HD on the employment and recreational lives of sixty nine patients and their partners of whom six individuals had an HD diagnosis. Fifty seven of this sample had moved from full time work to being unemployed and half the number of carers had to reduce their working hours in order to care for their partners. In well over half the sample these changes were viewed negatively with partners and patients referring to frustration, devastation, financial strain, social isolation, depression, disappointment, anger and grief.

Chapter 2. Literature Review.

2.1 Design and execution of the Literature Review.

The literature for this review has been assembled in two ways. There has been an incremental accumulation of relevant texts over the course of the research. In addition a systematic review was undertaken searching the three databases-Delphis, PsycINFO and Cinahl in the autumn of 2016. The following keywords were used in various combinations with each of the three databases-Huntington's disease; Qualitative research; Early onset dementia; Couple/Couple functioning/couple problems/family; Carer(s)/caregiver(s)/family member; Parenting; Diagnosis/impact of diagnosis/psychological impact; Testing/genetic testing; Sexual problems/sex; Suicide/suicidality/prevalence; Social stigma; Anger/reactive anger/anger and aggression; What helps/therapy.

Inclusion/Exclusion limits. No time limits were set and all online full text documents were considered. Only texts in English were subsequently considered for detailed review.

This search produced 136 references which divided under the following broad headings:-

Carers/caregivers (24), Child care (3), Couples (10), Depression (3),

Early onset dementia (5), Family life (2), HD (13), Professional involvement (14),

Quality of life (10), Research methods (4), Sex/sexuality/problems (7),

Stigma (3), Testing (15), What helps (2), Diagnosis (8), Suicide/suicidality (13)

The review that follows, and the earlier outline of HD, are based on a selection of sixty eight relevant texts from both the accumulated list of references and the more recent literature search.

In making this selection priority was given to papers which:-

Utilised qualitative research methods.

Described the prevalence of HD and other related illnesses.

Described the illness trajectory of HD.

Described the interactions between HD and couple/ family processes.

Located HD within a social and historical context.

2.2 HD and the impact on couple relationships

Reflecting on the range of papers that contribute to an understanding of the interaction between couple functioning and HD, it seems appropriate to consider what is meant in this context by a couple.

The idea of a couple is an elusive one partly because it refers to a relationship between two people which varies over time and about which opinions may differ. For example, one of the consistent findings from the literature is that couples contending with HD differ over an understanding of the disease's impact and severity. Kaptein et al. (2007) investigated fifty one couples having to manage HD with a view to exploring their individual perceptions of the disease and whether their perceptions are important in shaping their quality of life. Carers, as compared to their partners, identified significantly more HD symptoms and felt that their partners significantly overestimated the extent to which they were in control of their symptoms. These different evaluations of disease severity paralleled differences in the perceived quality of life with carers rating their quality of life lower than the ratings of their HD partner.

Moreover, there is no physical embodiment of 'coupledom' in the way that, say, 'carer' is embodied in a particular individual or a 'disease' is embodied in the malfunctioning of a human body. It is an abstraction or construction which seeks to capture how two people relate to one another over time and for various purposes. These purposes include:-

- The provision of an attachment figure for each other (Bowlby 1982; Fonagy et al. 2004).
- The procreation, education and socialisation of children, including the maintenance of moral values within a family and community.
- The establishment of an economic unit that generates income and assets, principally through paid employment.

The importance of these functions varies in significance according to stages in the family life cycle and, at any one time, we know that these functions can be interrelated; for example, the welfare of children is inextricably bound up with the stability of their parents' relationship and their economic status (Harold and Leve 2012). In this connection it is important to note

that the emergence of HD symptoms tends to be in mid-life when couples are very likely to have dependent children at home.

These comments are made to highlight and acknowledge the complexity of what ‘couple relationship’ means and to suggest that research studies are likely to provide windows of understanding into this large and complex phenomenon.

In fact, the literature that describes the impact of HD on couple relationships is small when compared to other progressive neurological conditions and diverse reflecting the multi-faceted nature and functions of adult partnerships, the different research questions and methods used to explore this complexity and the social and cultural context in which this work has evolved. The results from the literature search undertaken for this study has not identified a review of research focused on this particular area. However two reviews were found which are relevant. The first by Ablitt et al. (2009) looked at published work concerning the influence of relationship factors on living with all dementias. The second by Domaradzki (2015) looked at the impact of HD on the experience of carers. Both are relevant but Ablitt’s focus on all dementias does not account for the specific and unique challenges posed by HD which are lost in the broad sweep of the review’s remit and it does not take account of published work since 2009. The review by Domaradzki is relevant but by only focusing on the experience of carers in HD partnerships fails to take account of more interactive elements in the relationship between partners.

This section of the review will offer a resume of Ablitt’s paper because it offers an historical and theoretical framework for understanding the values and theoretical orientations that have influenced research into the links between dementia and couple relationships over the last 40-50 years. Additional references are added in when these amplify the points being made. The review will then continue by focusing on HD and linking research to different stages of the HD cycle.

Ablitt’s review claims that research in this area can be viewed from two perspectives-the historical and the theoretical. Taking the historical approach first, Ablitt identifies three stages covering the last 40-50 years. The first stage focused mainly on ‘carers’ and this ran from the early 1970s through to the late 1980s and 1990s. Cheston and Bender (1999) agree with the identification of this stage which they link to the introduction of Alzheimer’s disease

into the Diagnostic and Statistical Manual (111) in 1970 and the increasing medicalization of dementia from this time. In this process they argue that the lived experiences of people with dementia were disregarded and, as a result, they were not considered capable of 'being in a relationship'. One example of studies that might have reinforced this view were those that advanced the idea that dementia, in its advanced stages, could be experienced as a type of social death (Gilhooley et al. 1994; Sweeting and Gilhooley 1997), an idea that had three components:-

1. Anticipation of the patient's death current in the mind of the patient and those close to him, including professional helpers.
2. The patient suffers a progressive lack of awareness of and response to his or her environment; mutual recognition between patient and carer breaks down.
3. The carer/family acts on the belief that the patient is in some ways already dead.

While the evidence in these papers confirms social death as an important aspect of couple interaction, taken on its own, it does not address the qualities of the ongoing relationship from which it is drawn. At the time these ideas were current, the focus at both social and research levels was on healthy 'carers', a term which has no obvious pairing in the English language and which implies that care and support travels in one direction only. Both Ablitt and Cheston point out that the medical model of dementia began to be challenged in the 1980s and 1990s and this heralded Ablitt's second stage when research shifted to consider the experience of those suffering from dementia. This change in emphasis is particularly associated with the work of Kitwood who put forward a psycho-social theory of dementia in which the individual patient's 'personhood' is centre stage. Kitwood argued that 'personhood' develops and is affirmed through relationships with others and is, therefore, at odds with a purely medical model of dementia which tends towards a reductive biological formulation of dementia (Kitwood 1997).

According to Ablitt there then followed a third stage where researchers have attempted, more recently, to bring these two strands together and to focus increasingly on the interactions between the person with the illness and their partners. How the relationships between these two strands have been unpicked constitutes the theoretical perspective in Ablitt's review. Some of the work has sought to establish causal links so that one approach has been to see how the illness has impacted on the couple relationship. Those researchers who have adopted this approach have charted the negative impact of dementia on the quality of adult

relationships in a number of key areas-intimacy, reciprocity, communication, happiness and the overall quality of life. Ablitt notes that in these studies there is consistent evidence that the person with dementia, as compared with their partner, holds a more positive perception of the overall quality of the partnership, a point referred to earlier in relationship to HD (Kaptein et al. 2007). While Ablitt records overall negative impacts of dementia on couple functioning, the work of Clare (2002) and Baikie (Baikie 2002) offer a more mixed picture.

Clare interviewed couples who were in the early stages of managing Alzheimer's disease and identified two broad approaches to the challenges posed by the diagnosis. The first approach, and the one that most couples adopted, was a self-protective strategy which comprised efforts to hold on to pre-diagnosis ways of being and/or compensating for the limitations the illness imposed. A minority of couples adopted a second approach which is similar to the 'quest' narrative described by Frank (2013). These couples sought to integrate the illness into their lives by both fighting and confronting it and, thereby, seeking to come to terms with it. Baikie's study reinforced this view (Baikie 2002). She looked at couples where patients with mild to moderate dementia were aged between 60 and 69 years. She found that most partners maintained a positive pre-morbid image of their partner. In her sample she found that a clear diagnosis of dementia could, in fact, help the relationship by offering an explanation for behaviours and interpersonal difficulties that hitherto had evoked misunderstanding and blame. She did however note that in the early stages post diagnosis sexual difficulties emerged.

The second approach identified by Ablitt has been to assess how the quality of a couple relationship has impacted on the experience of living with HD. Research in this vein has produced consistent findings that lower levels of current relationship quality relate to increased depression in carers and people with dementia and to increased strain in carers. Lower levels of relationship quality are also said to correlate with reduced perceived self-efficacy in carers and reduced functional ability in people with dementia. Although higher relationship quality seems to be a protective factor for many, there is some evidence that higher levels of relationship quality can also be associated with later distress for carers suggesting that problems in coming to terms with the loss of a well-functioning relationship can, in itself, result in distress. This raises the possibility that how transitions are managed has to be considered in parallel with the changes themselves.

In both lines of research, Ablitt remarks that alongside the losses experienced by carer and partner, some positive aspects of the relationship may remain intact. In this connection, love, emotional warmth, affection and closeness of a non-intimate kind are mentioned as surviving under difficult circumstances. This points to the many layered and, perhaps, paradoxical responses to difficult circumstances.

Recognising the difficulties of establishing causal links in this area, another approach identified by Ablitt has been to describe frequently occurring patterns of interaction between carer and affected partner and the review describes four distinct shared dynamic patterns within the literature. The first, 'Continuity', refers to those couples who invest in maintaining the past identity of the person with dementia which has parallels with the first pattern identified by Clare. These couples operate on an optimistic view of the illness and their circumstances and the couple continue to enjoy companionship. These couples may underestimate the seriousness of the illness and find problems arising when forced to confront limitations imposed by the illness. The second pattern, 'Reciprocation', refers to couples where the carer acknowledges the changes brought about by the illness but is motivated to care because they have received care themselves from this person in the past or they believe that he or she would have done the same had the positions been reversed. The third pattern, 'Detachment' captures a type of relationship where there is little personal or emotional connection shown in the process of caring. It is suggested that for these couples the acknowledgement of feeling close is too threatening. The final pattern, 'Duty', refers to couples where the dominant sentiment behind caring is a duty or moral obligation; there is minimal mutuality in the relationship and the carer can be vulnerable to high levels of stress and then feel overwhelmed. This typology is interesting because it suggests that patterns of relating may be shaped by a complex mix of psychological defences and moral values. Although Ablitt's review records the move towards a focus on the relationship, it is noted that there remains a concentration on the experience of carers and a reluctance to engage directly with the person experiencing the illness, a point also made by Kitwood and Bredin (1992). Several reasons are put forward for this avoidance and these include ethical concerns that closer research involvement with the person with dementia will do harm, that there may be practical reasons making it difficult to access those affected and that there may be difficulties for researchers in engaging with behaviour that is emotionally challenging, difficult or disturbing. All these factors work against forming a balanced view of couple functioning as is clear from the proposed typology which emphasises patterns of caring behaviour.

2.3 Dementia and attachment styles

The onset of HD and other dementias results in major changes to the ways partners relate to one another. Significant change can elicit anxiety for one or both partners and attachment theory (and the classifications of attachment styles that flow from it) provides an important way of understanding the genesis of that anxiety, the different ways that anxiety is processed and the consequences of these differences for a range of measures including quality of life, neuro-psychiatric symptoms, carer burden, and emotional regulation.

Nelis et al (2014) undertook a systematic review of the literature concerning people with dementia and their carers. This study focused on two themes. The first was to examine how attachment ratings linked with quality of life, the expression of emotion and dementia outcomes. The second aim was to explore links between attachment ratings and the experience of carers. The findings from this review concluded:-

- That as dementia progressed and memory impairment increased, people with dementia in residential settings became less emotionally connected to partners and family members and could develop ‘parent fixation’ which was construed as a form of symbolic attachment. The sense of losing a connection with a partner sounds like the precursor to what has been referred to earlier as the later stage of ‘social death’.
- For patients with mid-to late stage dementia, secure attachment was related to more positive affect, better emotional regulation, greater sociability, less anger, fear and shyness. By contrast, those rated as dismissing of attachment showed an increase of behavioural-psychological problems including social withdrawal, delusions, and day/night disturbances. Those rated as ambivalent/preoccupied had higher levels of depression and anxiety.
- That carers, who were assessed as having an insecure attachment rating, experienced a higher burden of care.

The evidence from the Nelis review and other studies about the interaction of the respective attachment styles of both carer and care recipient is complex. In an earlier study by the same authors, Nelis et al (2012), looked at a sample of 97 couples where one partner had a dementia. This study found that while attachment ratings might be important for the psychological well being of the person with dementia and carer there was no evidence of

the attachment rating of one partner's attachment style impacting on the well being of the other and vice versa. Where there was a significant interactive effect, it arose from the neuro-psychiatric symptoms and behavioural problems of the person with dementia creating high levels of stress for the carer.

A different emphasis emerged from the study undertaken over two years by Perren et al (2007) to examine the impact of attachment styles on the behaviour of both the person with dementia and the well being of carers. This study drew on a sample of 116 married couples who were assessed at three stages across the two year period. It was found that :-

- The dismissing attachment style of the caregiver predicted levels of agitation and depression in the care recipient. It is not clear from the analysis whether increased levels of agitation and depression amounted to a changed attachment rating for the care recipient.
- A gender difference emerged; it was found that male caregivers were more likely to be securely attached and showed higher levels of anxiety at losing their partners than did their female equivalents, one explanation being that men stood to lose not only their partner but the social network she is likely to have built and maintained.
- Care giver avoidant behaviour and care recipients' insecure attachment style were associated with increased levels of dementia related problem behaviour.

2.4 HD and couple functioning during the life cycle of the disease.

2.4.1 Early days: when is HD recognised as a family or couple problem?

Individuals come to understand the presence of HD in their lives by different routes and manage this knowledge in complex ways.

Etchegary (2006) undertook a study seeking to understand how and when at risk persons first discover their family history of HD. Drawing upon twenty four semi-structured interviews with at risk persons and their family members, she explored when and how HD was discovered in the family. Qualitative data analysis revealed four different, though sometimes related, trajectories of discovery: (1) 'Something is wrong'. This route refers to the experience where family members may notice a worrying behaviour and this initiates a search which often results in various medical consultations before a firm diagnosis is made

(2) 'Out of the blue'. This refers to families where awareness of HD comes as a complete shock as often happens when an HD diagnosis is confirmed on a death certificate (3) 'Knowing, but dismissing'. Some family members will live with a vague awareness of HD existing in the extended family but choose to ignore the possibility that it may have direct implications for themselves and 4) 'Growing up with HD' some family members grow up with an HD relative whose condition may or may not be named. Etchegary was struck by the fact that, for most of her respondents, there was no singular moment when a discovery of HD was made. Rather it was knowledge that gradually crystallised from a state of preferred ignorance.

More recently Forrest Keenan et al. (2013) have examined how individuals came to realise that HD was affecting their partner and that they were living with a history of inherited HD. The study looked at the experience of thirty nine partners of HD patients and the analysis revealed four ways in which knowledge about HD was managed. There could be marital secrets in which knowledge held by the affected partner was withheld from the other, so that when HD was confirmed it came as a shock and evoked considerable discord. A second pattern was one where a partner was alerted to the possibility of HD being present in the family history but it was not made explicit; there was a 'turning a blind eye' by both partners to what couldn't be acknowledged. A third pattern was where knowledge was gained by observing and seeing another family member suffering from the condition and the partner 'putting two and two together' and, finally, the author describes conditions of marital ignorance where the family history was not known about and awareness of it came as a sudden shock to both partners.

2.4.2 Attitudes towards and consequences of testing

The decision whether to proceed with a test for HD when you are 'at risk' is extremely difficult to make because the consequences either way are of great potential significance for the person concerned and for other family members. It is then perhaps not surprising to learn that the proportion of at risk individuals who elect to go forward to being tested is as low as 25% (Forrest Keenan, 2013) and that most diagnoses of HD are made on the basis of clinical signs.

Oster et al. (2008) addressed this low take up by conducting a prospective longitudinal study of one thousand and one individuals in North America with a view to establishing their reasons for getting tested or rejecting the option. The principal reason for remaining untested was a comfort in living with risk and uncertainty and a view that, having been tested, it is then impossible to 'undo' knowledge. For those going ahead with testing their greatest concern was the possibility of losing health insurance as a consequence, a fear which was, in part, confirmed by the research by Williams (2010) mentioned earlier.

The results from testing can have huge significance for the person tested and for other close relatives. Richards and Williams (2004) undertook a qualitative study with the aim of assessing the impact of testing on couple relationships. Fourteen couples were interviewed where a partner was at risk of HD and the fourteen comprised nine couples who went ahead with the test and five couples who did not. Perhaps surprisingly, most couples who took the test said that the results of the predictive test did not adversely affect their relationship, a finding similar to that of Decruyenaere et al. (2004) who found that 70% of couples in his study were not adversely affected in their relationship 5 years later. Forrest Keenan et al. (2013) reported on similar adjustments after six months but found that, where individuals do get tested and receive a positive result, relationships will have to manage initial feelings of grief and shock. Taken together these findings suggest that couple relationships can be robust in the face of difficult news even if the implications of a positive HD test are serious.

However, Richards and Williams (2004) found that two couples who separated after a partner received a negative test result attributed the breakdown of their relationship to having had to live for years with the anxiety of developing HD. It seems that for these two couples it was the dispelling of the anxiety that triggered separation suggesting that this may be another example where difficulties in coming to terms with a new couple narrative may account for the breakdown. Moreover, two couples stayed together after a positive test result and attributed the shared loyalty and commitment to each other as the main factor in accounting for the resilience of their partnership. Richards and Williams conclude that these results point to the individuality and complexity of the psychological effects of testing, a point also made by Fowler (2000).

One way of illustrating the individuality and complexity of the responses to testing is to examine the experiences of one family. Anderson et al.(2016) offer a case study of a couple

in the late thirties with an eight year old daughter covering the three year period following the wife's predictive test which proved positive. The case study tracks a very tumultuous period for the couple during which the stress of the positive result resulted in the wife having an affair with a previous boyfriend and going to live with him in the hope of recapturing the person she was before testing positive, of the wife making a suicide attempt and eventually the couple divorcing. The authors argue that this couple's experience highlights the potentially devastating consequences of a predictive test and they argue for psychiatric follow up for some individuals.

2.4.3 Living with HD after diagnosis-the carer's perspective

As noted earlier there has been far greater attention paid to the experience of carers than to the adult partnership understood as a reciprocal relationship in which the separate and shared experiences of both partners are given equal consideration. This imbalance is reflected in the number of references generated in the literature search.

Domaradzki (2015) has produced a recent review of the literature on the impact of HD on carers. He reviewed thirty one papers of which fifteen were qualitative in orientation and sixteen quantitative and, from this second group, he identified six stress areas. These were 'Carer Overload' (problems in adjusting to role changes within the partnership, a conflict of roles between looking after an ill partner, attending to child care matters and managing the household, and a sense of losing a former quality of life, threats to personal identity, financial wellbeing and independence), 'Progression of the disease' (the increases strains that went with health worsening over long periods of time), 'Concerns over inheritance of the disease' (a reference to the anxieties parents feel in passing on the disease), 'Loss of meaningful relationships with partners', 'Negative impacts in the wider family system', and 'Strategies needed to cope'. These findings broadly overlap with Aubeeluck et al. (2012) earlier study where she and colleagues invited forty seven carers who were attending an HDA conference, to take part in six parallel small focus groups to explore the impacts of the disease on them. Four super ordinate themes emerged from their analysis of the group discussions. The first addressed being let down by the social and medical system, the second covered difficulties in the role of carer and the heavy burden this entailed. The third area addressed practical difficulties encountered in the caring role and the final theme covered the emotional difficulties carers faced, particularly in coming to terms with the loss of the pre-existing relationship with their partner. Under this final heading it was noted that some carers talked

of greater family cohesion being mustered in the face of daunting challenges echoing the findings of Ablitt and Richards and Williams mentioned before.

2.4.4 Living with HD after diagnosis-the couple's perspective.

In this section papers are described that either seek to capture the experience of both partners over time or which seek to address dynamic issues related to couples living with the reality or threat of a degenerative and fatal illness.

A starting point are the observations of 'at risk' asymptomatic adult children asked to reflect and record their memories of growing up in HD households. Vamos et al. (2007) sent questionnaires to forty asymptomatic adults between the ages of eighteen and thirty five years. Low family cohesiveness, high levels of family conflict and 'disenfranchised grief' (by which was meant having to mourn a variety of losses for which there is little or no public recognition or social support) were three of the significant findings emerging from their responses.

The image of communication breaking down between family members or parents existing on a conflictual basis invites a consideration of those papers that focus on partner interaction. Some communication problems are the result of physical impairments which are also likely to have reactive emotional consequences. Hartelius et al. (2010) looked at patterns of communication between close family members from the perspective of speech and language specialists. This was an in depth study involving eleven persons with HD, ten carers and seven family members. Overall they noted that patterns of communication had changed significantly and their analysis identifies important themes from the standpoints of the person with HD, carers and family members. Persons with HD talked of their thought and speech slowing down, of communicating being strenuous, of listening requiring a lot of concentration, of rich language being lost and, sometimes, realising that what is said was not intended. They often felt it difficult to understand others and to be understood themselves. Carers recognised the difficulties of their HD partners to express themselves, of finding them both getting stuck on particular topics and aware that these problems are heightened when their partner is depressed or stressed.

Exploring what created the greatest stress for patients and carers, Ready et al. (2008) found that functional and cognitive impairments are the greatest stressors but they also discovered that neuro-psychiatric symptoms affected carers and patients differently. There was evidence that it may be easier for the carer to cope with apathy than it is for the patient. Conversely the patient's aggressive behaviours were less troublesome for the patient than they were for the carer.

The significance of cognitive and personality changes brought about by HD in causing stressed couple relationships is described in the study by O'Connor et al. (2008). In their study there were four hundred and twenty three patients and three hundred and thirty five carers contending with motor neurone disease (MND), Huntington's disease (HD), Parkinson's disease, and multiple sclerosis (MS). Their analysis suggested that the overall predictor of marital satisfaction in people living with a neurological illness is social support satisfaction rather than the severity or length of impairments. However, patients and carers with HD had a significantly lower level of relationship satisfaction and sex life satisfaction than the other three illness groups which the researchers attributed to the cognitive and personality changes brought about by the disease plus the issues associated with the heritability of the illness. Further, patients with HD indicated a significantly higher level of relationship satisfaction than their carers, echoing the findings of Ablitt and Kapstein mentioned before.

Sexual problems alluded to by O'Connor et al. are confirmed by many other studies. Fedoroff et al. (1994) studied thirty two HD patients and their partners aged between twenty four and sixty five years and showed that 84% of HD patients and 64% of their partners had one or more sexual disorders. While there was a lessening of sexual activity between partners, there was a high incidence of increased sexual interest (or paraphilias) among the persons with HD. Reininghaus and Lackner (2015) replicate these findings and Kolenc et al. (2015) note that reduced sexual life between partners runs in parallel with advancing chorea and overall patient decline. Baikie in her study mentioned earlier (Baikie 2002) noted that a couple's sexual relationship deteriorated in the period following a diagnosis of dementia.

2.4.5 Couple resilience and what helps.

In this review there have been references to the fact that alongside the stresses, challenges and, sometimes, destructive consequences of HD, many couples readjust their expectations

and find within their own resources and between them the strength to find fulfilment and happiness in their lives together. Etchegary (2011) reflecting on research studies about life with chronic illness suggests that concepts of suffering and loss were prominent themes in the early years (e.g. the concept of social death) but that more recent work recognises that emotional strength and positivity can co-exist with living with background risk. If this is so, what is it that enables individuals and couples to readjust their lives, to move through the trauma of a chaos narrative and find a more balanced 'quest' narrative (Frank 2013) where the realities of the illness are not completely denied and can co-exist with making the most of life's opportunities?

There have been two ways of answering this question. The first has been to establish studies which have sought to identify couples who show higher and lower quality of life scores and compare the two in various ways. The second has been to examine some of the psychological mechanisms or processes that enable individuals within partnerships to manage anxiety and ill health.

The study by McCabe and O'Connor (2012) falls into the first category. They explored the question of couple resilience in the face of neurological illness. They divided a group of couples contending with a HD, MS, Parkinson's disease and MND into two groups of fifteen each- a high resilience group and a low resilience group in order to tease out the differences. While social supports-family, friends and partners- were cited across both groups as the single most helpful element in coping with illness, the high adjustment group used social supports differently from the low resilience group. The high adjustment group were more likely to draw on social support to maintain a normal life and to take responsibility for doing so in a proactive way while the low adjustment group drew on social support to provide assistance with funding or services particularly around tasks of daily living in the home. The former group emphasised the value of gaining information from voluntary associations while the latter group emphasised the value of material services provided. This descriptive identification of factors associated with resilience begs a series of further questions about what factors determine membership of the two groups.

In the second category, there are two helpful papers which seek to identify the processes that enable individuals and couples to cope. The first is a paper by Scerri (2015) who interviewed eight carers of HD partners who were either at home or living in residential care. The paper

draws the distinction between the ‘direct constant presence’ of the disease (which was most acutely felt when struggling with the dependency needs of their partners and managing concerns about the hereditary impact of the disease within their families) and the ‘indirect constant presence’ of the disease (by which they meant difficulties arising from changes to employment routines, financial strains, the loss of social activities and strained relationships in general but, particularly, the occurrence of marital problems). This is a helpful paper because it suggests that there are degrees of anxiety which are experienced in different ways. The second paper, already mentioned, by Etchegary (2011) had discussed how anxiety is managed under the heading of ‘salience’. Her study explored how people live with the risk of HD. Using IPA as a research method, she was interested to examine how the experience or intensity of felt risk (salience) varies over time and how high salience impacts on the self. In the background to her study, she noted that established research suggest that there are two broad themes that illustrate living with chronic risk: (1) biographical disruption, including threats to self-identity and changes to relationships with others; and (2) zones of relevance, the conditions under which risk is or is not salient and the study looked in depth at the experience of twenty four people who represented ten different families affected by HD.

Her results were that, while most respondents reported that risk was always there in the back of the mind (similar to Scerri’s ‘indirect constant presence’) it was not always salient so that there is a difference between ‘being at risk of’ and ‘being affected by’. Most tried to ‘normalise ‘the situation, ‘to get on with life’. Salience varied over time resulting from the interaction of various factors which included:-stages in the family life course (e.g. salience will increase when contemplating having children), the family history of HD (e.g. salience may increase when approaching the age when a parent’s HD began to show itself), disease related events in the family (e.g. salience may increase upon receipt of an HD test result or death in the family), personal beliefs or hunches, unique life events (e.g. salience may increase when starting university, at the point of couple breakdown, or when noticing odd behaviours). When risk became salient, there was evidence of biographical disruption which manifested itself by individuals feeling their personal identity and their social relationships were under threat.

2.4.6 Reflections on the literature review.

2.4.6.1 Limitation of the review.

1. Couple relationships can be understood as open systems in the sense that how they are formed and evolve is shaped both by factors within the boundary surrounding the couple but also by factors impinging on the couple from without.

This review has concentrated on how certain individual and shared psychological factors influence couple functioning and has not considered sociological and cultural influences. For example, the influences of social attitudes, social class and family income on couples managing an early onset dementia have not been examined.

2. The review has excluded a consideration of service user literature. People with HD and their carers have produced their own accounts of living with the disease. These accounts have emerged in book form, newspaper and internet based formats.

3. The review has excluded a consideration of pamphlets and other written material produced by the Huntington Disease Association (HDA) some of which is relevant to the study.

2.4.6.2 Implications arising from the review

This review suggests a number of general points which support the case for undertaking the research described in this thesis.

1. There appears to be an acknowledgment that the greatest burden in caring for people with HD falls on the family and, in particular, on partners, yet research into how couples are affected by the disease and how they adapt is in its infancy. There is a substantial literature on the role of carers supporting chronic illness, relatively less on carers supporting partners with HD and hardly any on exploring how couples, understood as an interacting psychological system, interact with the illness.

2. Cultural values influence research preoccupations. It is apparent that from the 1970s through to present times there has been a developmental sequence from focusing on carers, to recognising the person with dementia and, now, to paying increasing attention to the patient and partner viewed as an interacting system.
3. There is an acknowledgment in the literature that even when attempts are made to focus on couples there is a tendency for a slippage to occur as a result of investigators not engaging directly with the individuals affected by degenerative neurological disease. The result is that a bias towards a focus on carers is maintained.
4. The medical model tends towards describing the average and most likely presentation of HD symptoms with the result that the unique and varied way that the disease affects individual families is overlooked. While small scale in-depth qualitative studies attempt to capture this variability and complexity, they often carry the qualification that the representativeness of their samples is difficult to be clear about because of a hidden HD population that has difficulty in acknowledging the illness and engaging with research. For both these reasons, understanding the ways in which couples are affected is likely to be limited and partial.
5. As a consequence of the disease being inherited, any focus on couples has to take account of wider family concerns. These concerns are both ‘vertical’ in the sense that couples will be aware that illness in their lives will have emotional consequences for relationships with parents and any children, and they may also be ‘horizontal’ in that there may be questions about informing relatives in any extended family.
6. How individuals, couples and families live with knowledge about the disease is a recurring theme in much of the research. Living with a realistic appreciation of what a positive diagnosis means, while, at the same time, trying to make the best of what life offers shines through as a recurring dilemma. In this context it is striking that research from the broad field of dementia studies and the narrower field of HD studies points to the consistent finding that carers hold a more negative outlook than their partners in terms of the evaluation of symptoms and their quality of life.

The case for a study exploring the impact of HD on couples was given further impetus by the launch in 2009 of the National Dementia Strategy, updated later that year by the Implementation Plan (Department of Health, 2009). This plan emphasised the need for the provision of good quality early diagnosis and intervention for all patients and their families and as part of achieving this aim the Implementation Plan recognised the need for an

informed and effective workforce. Knowledge about the impact of diagnoses on the relationships of patients and their partners can contribute towards an informed and effective workforce. These connections are well established in the counselling field. For example, Bohart et al.(2002) reviewed forty seven studies and concluded that the correlations between levels of empathy and positive counselling outcomes represented a medium to large effect size. Conversely, one of the characteristics most commonly associated with negative therapeutic outcomes is therapists' lack of understanding (Mohr 1995).

One of the ways in which professional helpers, whether counsellors or not, gain insight and understanding of their clients' experiences is through reading or hearing about in-depth suitably anonymised accounts of individual, couple and family experiences. There is a limited literature describing the 'lived experiences' of couples coping with early onset dementia; a literature which, if expanded, might help advance professional skills as the National Dementia Strategy sets out to do.

Chapter 3. Methodology and Method

3.1 Philosophy of method

In its earliest stage, the aim of this study was to gain an enriched understanding of how a diagnosis of early onset dementia (of which HD is one variant) affects adult couple relationships. In order to contextualize this study, this chapter considers how the ‘lived experiences’ of couples contending with a variety of dementias can be theorized and researched.

3.1.1 What is meant by ‘lived experience’?

‘Lived experience’ has been defined in many different ways which then have implications for the ways in which they are researched.

An inductive perspective

Phenomenology is one important body of theory encompassing an inductive perspective. Carel (2016) defines phenomenology as a theory which focuses on “the experiences of thinking, perceiving, and coming into contact with the world; how phenomena appear to consciousness....phenomenology examines the encounter between consciousness and the world” (p 20). It is a philosophical tradition principally associated with a group of continental philosophers of whom Husserl, Heidegger, Merleau-Ponty and Satre are four of its most important members.

There are a number of general points that can be made about this perspective and their bearing on the current study:-

- It is an approach that seeks to elicit the experience of individuals rather than couples, families or groups. Consciousness, thinking and perceiving are all the mental activities of individuals. While it is possible to imagine that two partners in a relationship might share a common perception, the route by which that judgment is made is by abstracting and generalizing from the separate experience of each individual.

- The focus on individual experience encourages an idiographic approach to understanding ‘lived experience’. Allan and Eatough (2016) state that “Interpretive Phenomenological Analysis(IPA) is ideographic, starting with a detailed analysis of a single interview until some degree of understanding or gestalt has been achieved” (p409). If this approach is extended to researching couples, a starting point would involve interviews with both partners whether seen separately, jointly or a combination of both.
- Smith et al (2009) suggest two approaches for moving from claims about a single case to more claims about a data set. These are the inductive and quasi-judicial. The first involves taking an initial hypothesis derived from a single case and then testing it against each case in turn. The second is derived from the practice of the law where single cases are written up and considered in relation to others.
- Phenomenology does not admit an unconscious dimension to human experience; the focus of attention is on an individual’s conscious awareness at any one time. Therefore, a psychodynamic view of couple interaction involving defences and unconscious anxieties would be incompatible with a phenomenological perspective.
- Phenomenology, according to Husserl, is a ‘presuppositionless science of consciousness’ (Moran 2000 p.126) which means the observer “stripping away distractions, habits, clichés of thought, presumptions and received ideas, in order to return to what he called ‘things in themselves’. We must fix our beady gaze on them and capture them exactly as they appear, rather than as we think they are supposed to be....This setting aside or ‘bracketing out’ of speculative add-ons, Husserl called epoché- a term borrowed from the Sceptics, who used it to mean a general suspension of judgement about the world” (Bakewell 2016 p. 40-41). This approach means that phenomenology emphasises an emergent, inductive, or ‘emic’ understanding of experience rather than one which is theory driven, deductive or ‘etic’. The adoption of an inductive stance where the observer sets aside conventional ways of understanding, allows the possibility that radically new ways of understanding the experience of others may emerge.
- Phenomenology downplays the significance of the mutual interaction between observer and observed because this will cloud an understanding of the other’s

experience. Thus the co-constructed meaning of experience is less significant as a focus of attention than that on individual consciousness.

A deductive perspective

This is the diametrically opposed view to the inductive perspective, in that it claims the ‘theoretical’ lens of the observer shapes the way the experience of others is understood. Sartre thought that this constrained the capacity of any one person to know another’s mind; the other’s experience was essentially unknowable, or ineffable, as it was hidden behind the screen of the observer’s perceptual filters (Sartre 2003). On this reckoning, the interpretation of another’s lived experience then becomes located as an interactive phenomenon. The emphasis on an interactive understanding of another’s ‘lived experience’ is to be contrasted with an inductive approach which seeks to minimize the role of an interactive dimension as the observer seeks to drill down to an understanding of the unique and idiosyncratic experience of the individual’s conscious take on the world.

The inevitability of seeing the experience of another through the observer’s perceptual lens might be thought of as a logical corollary of the phenomenological view of experience for, if it asserts that every individual has a unique way of perceiving the world, it follows that there are as many different understandings of the world as there are people in it. The experiences of observer and observed are necessarily idiosyncratic and can lead to the postmodern despair that there are no absolute or essential truths and that truth is always relative and mutable as it emerges from social interaction.

There are a number of general points that can be made about the deductive perspective and their bearing on the current study:-

- It is a perspective that admits a range of different theoretical perspectives requiring the observer/researcher to be as clear as possible about the interpretive lens being used.
- Reviews of the application of different theoretical perspective has led to the identification of Thematic Analysis (TA)- a theory neutral research method.

- Within TA, a theme captures a common, recurring pattern, clustered around a central organizing concept. Whereas IPA procedures help the researcher stay close to the data in order that features of consciousness can be identified, the procedures of TA help the researcher identify patterns across the entire data set. This process involves processes of abstraction and generalization. When researching a sample involving couples the processes of generalizing and abstraction are likely to operate at two levels. First, these processes will apply to the partners of any one relationship. Second the processes will apply to commonalities operating between different partnerships.
- TA involves what Creswell has described as a data analysis spiral (Creswell, 1998), Following a process of data familiarization, initial coding takes place across all the data items from which themes are generated. These themes may then be clustered according to further abstracted sub-divisions (<https://www.psych.auckland.ac.nz/en/about-our-research/research-groups/thematic-analysis/frequently-asked-questions>)
- The process of generalization and abstraction central to TA raises questions about the status of the themes that emerge. O'Neill (1998) observes that as the analysis produces increasing levels of abstraction and generalization there are gains and losses. There is a move away from lived experience (understood as consciously experienced) to general claims about what a group shares in common, claims with which members of the group may or may not concur. To the extent that the claims do not describe the conscious experience of research subjects, the research cannot claim to be phenomenologically based. Nonetheless, findings may address 'lived experience' from an observer's standpoint and, thereby, contribute to understanding.

3.2. Implications for the study

This study was acknowledged from the outset to be exploratory. No qualitative research method, apart from the idiographic case studies of couples thought about from a psychoanalytic perspective, has been developed to explore the lived experience of couples. As the foregoing discussion seeks to outline, there is a limitation on the capacity of IPA to move beyond the singular experience of one person; an exploration of couple functioning requires exploring, at least, what two people share and differ over so that some degree of abstraction and generalisation informed by, theory, is required.

Accordingly, it was decided that the study, which aimed to describe, understand and interpret the lived experiences of a small sample of couples where one partner had been diagnosed with an early onset dementia, would, following Holloway and Wheeler (2010), recognize that:-

1. The data have priority or primacy in a way that allows preconceived theory and assumptions to be challenged.
2. The data are context bound resulting in a tension between emergent, inductive or 'emic' knowledge and interpretations that, driven by the presuppositions of the researcher, are, therefore, theory driven, deductive or 'etic'.
3. Researchers immerse themselves, as far as is possible, in the experiences of the people whose behavior and thoughts they wish to explore. This is undertaken on a basis of respect for and equality with research participants.
4. Reflexivity, that is the thoughts, observations and feelings of the researcher generated by the research engagement, is a source of important research data. It follows that the relationship between researcher and research participants is likely to change and evolve over time as the researcher understands progressively more about the phenomena under scrutiny and develops his research interventions accordingly.

In order to meet these criteria a number of research methods were considered. These included Grounded Theory (Glaser and Strauss 1967; Charmaz 2011), Narrative Analysis (Andrews et al. 2008) and Interpretive Phenomenological Analysis or IPA (Smith et al. 2009). Even if some researchers are able to 'bracket off' in the way that the IPA method requires, in situations where the discussion of important family matters requires trust in the researcher, it seems reasonable to assume that the reality of the researcher-what he or she brings in terms of appearance, personal manner and ways of thinking-will influence the responses of research participants. Recognising the influence of the researcher, supports the idea that some elements of what is discussed in research interviews need to be thought about as being jointly constructed and unique to the research context.

From a review of a number of possible research methods it was decided to use a modification of the Free Association, Narrative and Interview Method (Hollway and Jefferson 2000) as this seemed to offer a recognition of the socially constructed nature of research data and also allowed for the researcher to employ a psychodynamic lens when interpreting data. FANIM seeks to understand the meaning attributed by individuals to certain sorts of experience,

particularly those that evoke anxiety. Hollway and Jefferson developed this method when exploring how people talked about and were experiencing the threat of crime in their daily lives.

It is based on a number of assumptions which emphasise the context specific meaning of research data and the significance of the researcher's interpretations in elucidating meaning. These include the following:-

1. 'We don't know ourselves', a psychodynamic view which claims that we have blind spots/defences. This claim challenges what they call the transparent self-problem i.e. a mistaken belief that we know ourselves and 'say it how it is'
2. Words do not have identical meanings across contexts and between individuals: a claim that challenges the transparent language problem i.e. a mistaken belief that words have rigidly consistent meanings regardless of context.
3. The data from research interviews is co-constructed. Minimally, the claim is that all data is interpreted data which implies that the contribution of the researcher must be taken into account or 'theorised'. This then creates a solipsistic system or circular problem in which the researcher is both observer and subject, sometimes referred to as the 'hermeneutic circle'.
4. The interpretation or understanding of what a person says has to take account of an interplay between what they bring to their understanding by virtue of past history (this is operationally defined as a psychodynamic formulation of how the individual manages anxiety-the defended self) and a set of publically defined meanings relating to the subject under discussion (for example, that dementia equates to social death). The product of this interplay produces what they term the psychosocial subject.

Two features of this method raised questions about whether it could be adapted to this project. The first, which applies to many qualitative methods developed for interviewing individuals, is whether it could be adapted to interviewing couples. The possibility of doing so raised the innovative prospect of developing the concept of a psychosocial couple where the couple is thought about as an entity in itself. If the essence of FANIM is to understand the interplay between an individual's internal world and a particular social discourse, the formulation of a psychosocial couple would require an understanding of the interplay of two persons' internal worlds interacting together with a particular social discourse.

The second problem is whether a psychoanalytic theory other than a Kleinian one (the theory used by Hollway and Jefferson) could be used to formulate the idea of a defended couple. Changes to relationships brought about by chronic long term illness often involve having to confront loss, separation and the reassignment of caring roles. These factors raised the possibility that the use of an attachment theory informed understanding of change might better fit participants' experience.

In applying attachment theory it was not envisaged that a formal attachment rating interview schedule, like the AAI, would be used. Rather, general ideas about attachment security and insecurity might be used to understand couple interaction.

Attachment theory is a spatial theory. The theory postulates that within all humans there is an autonomous attachment behavioural system that periodically seeks *proximity* to the major attachment figure when the relationship to that figure is broken or under threat (Bowlby 1982). Proximity is mediated by looking, hearing, talking and holding. When proximity is established the individual feels secure and is able to relax and to get on with daily living and, especially in children, to explore their environment. However not all children or adults have had good enough experiences that enable them to engage the attentions of their primary attachment figures and develop, instead, patterns of insecure attachment behaviour.

Main (1985) identified two broad categories of insecure attachment in adults. These are the dismissing adult and the preoccupied adult. The hallmark of the dismissing response is that the adult when faced by an attachment threat, typically a significant separation, downplays the emotional significance of the disrupted relationship. Phrases like 'putting one's head in the sand' or 'turning a blind eye' may be indicative of dismissing responses. By contrast the preoccupied adult, under similar conditions, remains caught up and finds it difficult to separate from the primary attachment figure. Anger is consciously denied and the individual clings, however ambivalently, to their attachment figure. Whereas dismissing adults may turn away both emotionally and physically from their partners, down playing their feelings, the reverse happens for pre-occupied individuals who stick adhesively, or cling, to their partners and up-regulate their emotions. Lyons-Ruth and Jacobvitz (1999) also identified a fourth sub category which could be applied to the three categories of secure, dismissing and pre-occupied. This is the sub category of unresolved which is identified by a person's intermittent tendency to talk in the present tense about past events and it is suggested that this arises as a result of an unresolved trauma from the past linked to a significant loss or experience of having been abused.

3.3 Advantages and Disadvantages of the interview method

There are advantages and disadvantages attached to using audio-recorded face-to-face interviews and ‘open-ended’ questioning techniques.

Advantages:-

- The principle advantage is that this approach offers the possibility of gaining rich, in depth data combining emotional depth with narrative content. This potential can also work against participants’ interests if the interview format heightens anxiety in unhelpful ways. This possibility raises ethical issues which are discussed later.

Disadvantages:-

- Roulston et al (2003) identify four challenges faced by novice interviewers. These are
a) unexpected participant behaviour; research subjects may have their own agendas which subvert research aims; when interviewing research subjects with chronic illnesses there may be physical and psychological impediments to verbal engagement.
b) consequences of the researcher’s own actions and subjectivities; the researcher may not act consistently from one interview to the next or may lose sight of the research focus,
c) phrasing questions; questions may not be put clearly enough and may not be sufficiently open and
b) dealing with sensitive issues; researchers may be thrown by dealing with emotionally challenging topics.
- There are particular challenges when interviewing couples. A choice has to be made about whether to interview couples separately, jointly or in a combination of both. Joint interviews tend to reveal a picture of current couple interaction but may also be experienced as too exposing. By contrast, separate interviews may feel more comfortable for participants but may also reveal couple confidences which create ethical problems for the researcher in how they are reported.

As a consequence of the disadvantages outlined above, other sources might have also been studied. These include:-

- Personal diaries.
- Internet based patient forums

- Live patient and carer groups such as those organised by the HDA.
- Service user autobiographical publications

3.4 Scoping Review

(See Appendix 1 for Ethics approval, Invitation Letter, Participant Information Sheet, Consent Form and Interview Schedule)

The NHS service, where it was hoped to base a future research project, required, as a matter of policy, that a Scoping Review be undertaken before any later proposal was considered. This was undertaken between autumn 2009 and spring 2011 and comprised two elements. The first, for which ethical approval from the Faculty was granted, involved a consultation exercise with service users. The second element, which ran in parallel with the first, involved consulting with colleagues who were either experienced in the field of couple therapy/research or/and had experience in working or living with chronic illness, about the proposed research.

3.4.1 Consultations with service users

The consultations with service users had three aims:-

1. To help formulate and refine specific research questions
2. To gain advice about how research results can be most usefully disseminated
3. To explore the possibility that service users might be involved in helping to analyse research data

3.4.1.1 Participants

It was proposed to interview up to a maximum of five couples recruited from service users attending an Alzheimer's Society Day Centre and from the membership of the Huntingdon's Disease Association (HDA), via its national newsletter.

3.4.1.2 Inclusion Criteria

Couples where one partner has been diagnosed with a young onset dementia, within the last three years and who, in the opinion of care staff can helpfully contribute to the scoping review without harm to themselves. The requirement that a diagnosis has been made within the previous three years makes it likely that patients will be in the early stages of their illness

when levels of cognitive, emotional and physical functioning will be adequate to engage with the demands of being interviewed.

3.4.1.3 Exclusion criteria

Patients without partners.

Patients who in the judgement of colleagues in the Alzheimer's Society and the HDA are too ill to participate.

Patients who are involved in another current qualitative study.

3.4.1.4 Recruitment process and ethical considerations

Two couples volunteered to be interviewed jointly. One couple was introduced personally at a day centre and the second couple made contact having seen the research advertised in the HDA Newsletter. In line with the research protocol, both couples were each sent a Participant Information Sheet and Consent Forms (see appendices for copies) prior to being interviewed in their homes. In recruiting couples, care was taken to ensure that consent to participate in the study was freely given and that provisions of the Mental Capacity Act (Mental Capacity Act 2005; Office of the Public Guardian 2009) were adhered to. An important requirement of the Act is that potential participants in research projects should be given enough time to fully consider their decision about taking part. This requirement is important for those potential consultees whose neurological problems may intermittently impair levels of attention, concentration and recall. Couples were given at least two weeks in which to consider future involvement with the study.

3.4.1.5 Consultation method including Reflexivity

The consultations were held in the homes of both couples. Interviews were audio recorded and were subsequently transcribed. No attempt was made to capture the speech impediments of the two speech impaired partners.

Diary notes were made before and after interviews. These summarised thoughts, reflections and observations of the consultation process and were later referred to in analysing interview content (Hammersley and Atkinson 1983; Smith 2008).

3.4.1.6 Brief out lines of both couples

[Care has been taken to ensure that the identities of participating individuals have been suitably disguised.]

Couple A

John, who was of pensionable age, was married to Ann, some 7 years younger who had been diagnosed with HD six years before I interviewed them. Keeping to the interview task was difficult for a variety of reasons. Ann's illness had resulted in a marked speech impairment coupled with a capacity to be preoccupied with certain themes. It was difficult to understand whether her repetitiveness was linked to a neurological problem and/or an insistence that, in spite of her communication difficulties, she was determined to get her points across.

Couple B.

I was introduced to Carol and David through my attendance at an Alzheimers Society Day Centre and, at our first informal meeting in a large group, Carol chose to speak with me out of David's hearing. She told me how dependent David has become on her and of how she manages by putting her 'head in the sand' and only thinking a few weeks ahead. She also told me then that this means of managing her current problems linked to a similar way she found to cope when abandoned by her mother as a child but she was now worried that this mechanism was not working as effectively as it once did.

Carol was just of pensionable age and David seven years younger. He had an Alzheimers type dementia diagnosed four years before meeting with them. David was chair bound and it was noticeable that, because he had a left sided weakness, he needed regular propping up with cushions on that side. A further consequence was that his head fell to the left and his left sided facial muscles were weakened resulting in him dribbling a lot. He talked with a low gruff voice that was, sometimes, difficult to hear and his verbal contributions were brief and, often, monosyllabic. Carol tended to speak for them both unless David felt an urgent need to make his point when he would do so clearly.

3.4.1.7 Results bearing on the interview schedule/research aims.

Both couple consultations demonstrated that seeking views about interview questions took place in a context shaped by illness related limitations and by the opportunities the interviews offered for the couples to pursue their own agendas. For both couples the well partner was the stronger voice with John often attempting to bring Ann back to focusing on questions asked and Carol compensating for David's limited capacity to speak clearly. From the beginning

arrangements for the interview were channelled through the well partner, an arrangement that set up an imbalanced relationship with them as couples. However, despite having the weaker voices, both ill partners conveyed an insistence on having their thoughts and feelings heard. Ann used the interview to convey two important messages. The first was a concern about her adult children, one of whom has been diagnosed with HD and was experiencing a cluster of psychological problems. The second was an insistence to convey a resilient and strong sense of self in the face of her illness. This took the form of describing the important job she had before her illness resulted in what, she believed, was a case of constructive dismissal and asserting her contributions to current domestic life, especially through cooking.

Echoing Ann's need to assert her sense of personal worth, David used the interview to remind Carol that before he became ill he had been her 'boss' at work where they had been colleagues. This statement of his former potency sat poignantly alongside his sadness that they no longer had an intimate sexual relationship and Carol's relief that this aspect of their relationship had ended. Their divergent views about the changes in their sexual relationship raised the possibility that exploring this aspect of the transition from a pre-illness relationship to a carer/cared for relationship might be important in the main study.

John and Carol, too, had important messages to convey about their respective relationships which signalled possible subjects for future attention. John, as the 'healthier' partner, struggled to balance feelings of concern for Ann with feelings of anger. Humorous and somewhat patronising references to keeping a watchful eye on her wellbeing-he referred to her a 'poor old thing' and 'acting like a 2/3 year old at times'-coexisted with explicit acknowledgement that he had problems in managing his anger; 'I lose my rag at times' and 'Perhaps I can be too protective'. Ann was fully aware of this difficulty but insisted that they remained a strong couple and continued to enjoy an intimate relationship. This aspect of their interaction raised the possibility that asking how frustrations and anger are managed might be significant in future interviews.

Carol had told me that she copes with her difficult predicament by putting her 'head in the sand' and putting up 'a brick wall' suggesting an avoidant/dismissing defence. This seemed reflected in her frequent use of the impersonal 'he' when referring to David, in his presence. Interestingly, she had agreed to the interview saying that her 'brick wall' was not as impermeable as it had been and this comment, plus her comments about how unsupported she felt, raised the possibility that she might have wanted a relationship that had a therapeutic purpose.

3.4.1.8 Results bearing on the development of FANIM.

Both couples received diagnoses longer ago than the specified 3 years mentioned as an inclusion factor in the Scoping Review proposal. This may have contributed to the difficulty of maintaining a focus on experiences around the diagnosis itself and of eliciting details about past history. For both couples their preoccupations were very much centred on their present circumstances which had understandably changed and deteriorated since the times of their diagnoses. This quality of concern in the immediate present evidenced in one-off joint interviews had strengths and weaknesses from a research perspective. The strength was that clear pictures were conveyed of how the dementias were impacting on current lives, the weakness was that it was difficult to gain full developmental histories of each partner as would be required when using FANIM. Asking detailed questions about the characteristics of their relationship twelve months either side of their diagnosis in order to understand how their partnership had changed did not work suggesting that, if more detailed histories of each partner was required, there might need to be individual in addition to joint interviews.

3.4.1.9 Results bearing on the analysis of data.

Both couples were pleased to help with the development of the study although in Carol's words it was possible to hear some ambivalence. She said she "wanted to help someone not go through so much anxiety or just inform how things might be, people do say how things might be but I think for me I put up a 'brick wall', well I am sure I did". It is possible to detect in this comment both a wish to help others but recognition that, for her, she has a characteristic way of avoiding what is difficult to contemplate. John was also explicit in wanting to share experiences with me and felt that others would be, too, because "Well, I think that these days people are more aware of HD" implying that he wished to contribute to this publicity. He felt that research findings could be usefully spoken about at the HDA national conference and at local branch meetings of the HDA

3.4.1.10 Results bearing on the dissemination of results.

Both couples gave qualified replies to my enquiry about whether they thought service users might be involved in helping to analyse research data. Carol said that she might like to help with analysing data "if asked"-although I had not spelt out what analysing data meant in precise terms. David suggested that a friend in their local HDA branch might be a good person to ask.

3.4.2 Consultations with colleagues

During the early part of 2010, a brief outline of research intentions and a draft of the Interview Schedule were circulated by email among colleagues who had expressed a willingness to comment on the research at its earliest stages. These colleagues included specialists in the practice of couple therapy, experienced researchers into couple functioning and colleagues with a special knowledge of dementia. Their main written comments were:-

- a) That by seeing couples in joint interviews only, some important data about the adaptations partners have to make would be lost. It was thought likely that important information would be disclosed in separate meetings that could not be shared in joint interviews. On the other hand, it was acknowledged that seeing partners separately can present ethical problems if 'secret' knowledge cannot be shared with a partner and/or published.
- b) That asking questions about the first twelve months of the relationship might not be helpful if the aim was to discover the emotional chemistry that linked partners together. If that information is needed it was suggested that the question be asked directly.
- c) That interviewing couples some distance in time from events is a positive advantage if a considered and balanced view is being sought. The experience with both couples seen in the Scoping Review was that there is no such thing as a considered and balanced thought if by this is meant that there is some settled view. Both couples had to appraise the past through the filter of changing and deteriorating circumstances.
- d) That edited words on a printed page cannot fully capture the struggle of someone trying to express themselves when their illness adversely affects their capacity to speak. Poor pronunciation and a competitive struggle to speak alongside a more able and articulate carer is very difficult to capture in the two dimensional medium of the printed page.
- e) That couples should be interviewed in their own homes rather than in an office because they had a better chance of 'holding their own' as a couple. A home setting was compared to a medical setting where the roles of carer and patient are ascribed in ways which can be limiting and inhibiting.
- f) That it would help if couples are asked about what makes them frightened and/or angry.

The analysis of the consultations with service-users and colleagues resulted in a report produced in April 2011 which formed the basis of discussions within the Faculty and with colleagues in the NHS.

A significant development at this point was to learn that the psychiatric service within the NHS where it had been planned for the study to be based was shut down and colleagues with whom tentative plans had been made were either reassigned responsibilities or left the service. This development meant that fresh negotiations with other parts of the NHS would need to have been started if the focus of the study was on a range of early onset dementias as had been intended. Moreover, at that stage, there were strong reasons to refocus the attention of the study on HD. While the link with the NHS was broken, that with the HDA had proved positive and the limited literature search undertaken at this stage, combined with advice from NHS colleagues, pointed to the relative dearth of research looking at families contending with HD. For these reasons it was decided to limit the research sample to couples where one of the partners had HD and to recruit the sample through the HDA. The viability of proceeding in this way was confirmed by the national office of the HDA offering their continuing support of the study and by discussing research plans with three Regional Coordinators of the HDA. They offered advice about the distribution of HDA branch organisations within their areas and contacts I might make at branch level in order to help recruitment to the study. They also agreed to offer advice about the suitability of couples being included in the study if it was thought involvement in research interviews would be emotionally destabilising. The outcome of the scoping review and subsequent discussions was a clarification of the research questions and research method to be used.

3.5 The Main Study

(See Appendix 2 for Ethics approval, Advertisement in HDA Newsletter, Invitation Letter, Participant Information Sheet, Consent Form, Individual Interview Schedule and Joint Interview Schedule)

3.5.1 Aims

The overall aim of the study was to explore how a diagnosis of HD impacts on couple relationships. In the process of pursuing this broad aim there were three subsidiary questions:-

1. Is it possible to think of a diagnosis of HD as an attachment threat?
2. How does an understanding of each partner's formative attachment experiences throw light on their reactions and adjustments to an attachment threat as presented by HD?

3. How feasible is it to modify FANIM so that it can be used to generate an understanding of couple interaction and behaviour from an attachment perspective

3.5.2 Method

From the Scoping Review the modifications to FANIM were:-

1. Couples would be first interviewed individually and, then, jointly within a two week period. The rationale for individual interviews was that these would provide a picture of each partner's developmental history. Joint interviews would provide a picture of couple interactions and responses to HD.
2. Interview Schedules for both individual and joint interviews would be informed by an attachment theory perspective. While an open ended approach to interviews would be adopted, direct questions and prompts, when needed, would seek to elicit information about how, in the past and present, significant personal relationships had been formed, changed and relinquished.

3.5.3 Participants, sample size and homogeneity

It was proposed to interview four couples in their own homes if possible. These couples would be recruited via the HDA newsletter or by hearing about the study from talks given by me at branch meetings of the HDA. Sample size was determined by the resources available to process data (four couples would generate between twelve and eighteen hours of interview data) and by advice from supervisors.

The sample would be self selected, subject to inclusion and exclusion criteria and , as such, would be a 'purposive' sample. Its relationship to the wider HD population in terms of demographic, socio-economic status and medical characteristics would not be known; the aim of the study did not include making generalisations from the sample to the wider HD community.

3.5.4 Inclusion criteria

It was intended to include couples where one of the partners had been formally diagnosed with HD between two and ten years earlier. This time range was chosen so that couples would have had time to make any adjustments to their relationship since the diagnosis and both partners should be able to reflect on their experience i.e. memories would not be seriously compromised.

3.5.5 Exclusion criteria

It was intended to exclude couples where there was judged to be a pre-existing level of behavioural and/or emotional disturbance that would make exploration of couple functioning potentially destabilising. This judgement was to be made initially on the basis of how the initiating partner, normally the caregiver, replied to questions about their partner's health and functioning. Should this discussion raise any doubts about the inclusion of a couple, further advice was to be sought from the Regional Care Organisers of the HDA.

3.5.6 Informed consent and Capacity

An important requirement of the Mental Capacity Act (Mental Capacity Act 2005; Office of the Public Guardian, 2009) is that potential participants in research projects should be given enough time to fully consider their decision about taking part and to offer their freely given consent. This requirement is important for those potential research subjects whose neurological problems may intermittently impair levels of attention, concentration and recall. To accommodate this consideration, the study adopted a minimum gap of two weeks between first discussing the project and holding the first interviews. During this period each couple would have sight of the Invitation letter, Participant Information Sheet and Consent Forms. It was anticipated that this would allow sufficient opportunity for both partners to discuss their participation and would allow any uncertainties or doubts to be addressed.

3.5.7 Anonymity

In the analysis and report writing stage of the study it was recognised that the anonymity of all participants needed to be ensured by removing or changing all identifying personal references. The standard adopted to secure this level of confidentiality would be that no outside third party beyond the research group and the participants themselves would be able to identify the transcribed and edited texts. This requirement is especially significant to honour because members of the HDA are often known to one another. For this reason the analysis of interview transcripts has not involved looking at how participants have related to the HDA. Detailed discussion of this relationship was felt to be too compromising of confidentiality particularly as all of the participants volunteered through membership of the HDA and many of them have asked to be informed of research findings.

3.5.8 Data Protection Act and storing data.

All documents containing identifying links to research participants have been stored according to University Regulations and the provisions of the Data Protection Act 1998. All hard copies of documents have been kept under secure and locked conditions. All electronic documents are password protected and anonymised. All research materials will be stored for ten years in accordance with University Regulations.

3.5.9 Relational ethics

While the information exchange prior to the interviews was intended to cover all predictable implications of participation in the study, it was acknowledged that the use of open ended questioning in relation to a debilitating and degenerating neurological condition would touch on sensitive and emotionally charged subjects which might be experienced as welcome by some couples but destabilising for others.

Hollway and Jefferson (2000) comment that in these circumstances a distinction needs to be drawn between becoming distressed and experiencing harm. It is possible that participants may become distressed during interviews and it would be very surprising if this did not happen. However, becoming distressed about distress-inducing circumstances is not in itself the same as being harmed. They argue that two crucial points follow from this:-

- Giving consent cannot be a one-off event, limited to signing the Consent Form. Participants need to be reassured that they can withdraw consent at any time. It was planned that this would be made clear to all participants and that their experience would not then form part of the analysis.
- The interviewer needs to remain attuned to what participants are feeling and to be respectful of what they can manage in terms of contributing to the interview task. The exploration of personally sensitive issues needs to be done in a thoughtful and kind way. To help mitigate the unsettling potential of these interviews it was decided to include the provision of a 'wind down' discussion after the audio equipment was turned off, something that proved helpful for the couples seen in the Scoping Review.

3.5.10 The recruitment process.

Ethical approval for the study was granted in January 2014 and the invitation for couples to participate in the study was included in an HDA Newsletter as a result of which two couples replied and interviews with both followed in February and March of that year. The first couple completed the full set of interviews-two individual interviews followed by a joint interview- but the second couple withdrew from the process following two individual interviews in their home. Preparations for their interviews had not involved the female partner reading the Participant Information Sheet and Consent Form which had been forwarded to her male partner who had set up the interviews. Why the forms had not been read was not made clear when we met for the individual interviews but the subsequent refusal to agree to a joint meeting suggested that, among other possible explanations, the failure by the couple to agree participation in the study played a part. The couple chose not to reply to follow up correspondence and their 'de facto' withdrawal means that material from their individual interviews cannot be included in the research data set.

There then followed a period of twelve months when no further couples came forward despite efforts to promote the study. These initiatives included a visit to a local HDA branch meeting, liaison with the Regional Care Coordinators of the HDA, a follow up entry in the HDA Newsletter, publicity for the research displayed at a study day in September for professionals and inclusion of a flyer about the study in the delegates' packs for that year's Annual Conference which I also attended. At the branch meeting, two male carers indicated that they would be willing to be interviewed but were sure that their partners would refuse. A similar message came across in informal discussions with carers at the AGM and Conference, several of whom attended without their partners who were said to be either too ill to attend or were described as denying that they were ill and wanted no part in an activity that questioned that belief. One outcome of the conference was a further visit to give a talk to an HDA branch meeting.

In the spring of 2015, it was decided that, as a consequence of the difficulty in recruiting couples, and the possibility that more success might be achieved by interviewing one partner of a couple, the research sample was extended (See Appendix 3 for Ethical approval). In

addition to the hope that three further couples would be recruited, ethical clearance was granted to interview four individuals without their partners. In all other respects the aims of the study including inclusion/exclusion factors remained the same. An announcement of this change was made through the HDA Newsletter and by giving talks at four HDA branch meetings. As a result three individuals and one couple offered to be interviewed and their interviews were all completed in the autumn of 2015. One of the individuals who volunteered was keen for me to interview her partner whose HD required him to be resident in specialised accommodation near to the family home. He had indicated to his wife that he was prepared to see me and this resulted in an unplanned extra individual interview of about 20 minutes duration in his residential care home setting. In the analysis that follows this interview is not included.

The research sample does not conform to the criteria specified in the research proposals in two important respects. First, the sample numbers anticipated for couples was not met. In the main study it was planned to interview four couples. Only two couples completed the full programme of two individual interviews followed by a joint interview which limited the opportunity to test the viability of a modified FANIM for interviewing couples'

Second, it was planned to interview couples and individuals where a diagnosis of HD was made no more recently than two years and no longer than ten years beforehand. This time window was chosen because it was thought that it would give a picture of adjustments to the illness post diagnosis and, in relation to couples, would enable both partners to be involved. In fact, in the two instances where couples completed the full set of interviews, one couple, had received a diagnosis seven years before the interviews while another couple received a diagnosis nineteen years before. This much longer time gap did not compromise the ability of this couple to engage fully with the research task.

Of the four individuals who came forward to be interviewed diagnoses were made in a range extending from twenty months to twenty years before they were interviewed. Difficulties in the recruitment of couples and the value or otherwise of specifying a time window for when the diagnosis was made will be discussed later in this report.

3.5.11 Data Analysis

This study adopted the semi-structured interview method used in FANIM and meets the aim of a qualitative research method to elucidate the subjective experience of participants.

All interviews were scheduled for a maximum of ninety minutes and were audiotaped on two machines. The amount of interview material in the autumn of 2015 required the help of an approved professional transcriber for seven interviews. Transcriptions did not attempt to capture idiosyncratic mannerisms and the texts can be described as ‘cleaned up’ speech.

It was proposed that the analysis of interview content would involve a thematic analysis (Braun and Clarke 2006) at two levels. The analysis would allow the identification of emergent themes i.e. those not predicted by the theoretical categories underlying the interview schedules, but the analysis would also seek to identify themes within an attachment framework. Therefore, the analysis would seek to identify ‘bottom up’ (emergent) and ‘top down’ (attachment theory driven) themes.

Phases of Thematic Analysis. (from Braun & Clarke, 2006)

Phase	Description of the process
1. Familiarising yourself with your data:	Transcribing data (if necessary), reading and re-reading the data, noting down initial ideas.
2. Generating initial codes:	Coding interesting features of the data in a systematic fashion across the entire data set, collating data relevant to each code.
3. Searching for themes:	Collating codes into potential themes, gathering all data relevant to each potential theme.
4. Reviewing themes:	Checking in the themes work in relation to the coded extracts (Level 1) and the entire data set (Level 2), generating a thematic ‘map’ of the analysis.
5. Defining and naming themes:	Ongoing analysis to refine the specifics of each

	theme, and the overall story the analysis tells; generating clear definitions and names for each theme.
6. Producing the report:	The final opportunity for analysis. Selection of vivid, compelling extract examples, final analysis of selected extracts, relating back of the analysis to the research question and literature, producing a scholarly report of the analysis.

For the purposes of understanding a social discourse about HD, it was planned to review the publications issued by the HDA. It was assumed that their range of publications, providing information and advice about managing HD, would contain important socially constructed assumptions about the disease.

3.5.12 Management and analysis of data.

Managing and analysing seventeen hours of transcribed interviews was a complex task arising from the amount of data and the fact that, in three way meetings, there were three voices to track. Familiarisation with the data was also complicated by the pace at which the interviews accumulated and by the use of an approved transcriber for seven of the eleven interviews. I transcribed the first interview in the spring of 2011, a further three interviews in the spring of 2014 and there was then a gap before the final cluster of seven interviews were completed and professionally transcribed in the autumn of 2015.

My familiarization with the data was progressive over time with my understanding being cumulative and uneven-I was aware that my understanding of the interviews I transcribed was qualitatively different to those for which I obtained outside help. I tried to accommodate this difference by giving extra time to reading the professionally transcribed texts.

Following close and repeated reading of the transcribed interviews, I experimented with different forms of text layout. I settled for a two stage process.

The first stage involved extracting codes from the data. These were inserted in the text in a sequential manner with codes for participant contributions and codes for interviewer contributions typed in different colours. As an example of this process I give a brief extract from a joint interview with Couple 3 (see later). It is an excerpt from the start of a joint interview where the couple are commenting on whether they had discussed together the contents of their individual interviews with me. First stage codes are underlined.

P8M We did not discuss the answers that we gave you between us at all.

G making clear that what was said in the individual interviews was not shared between them

P8F No

P8M What we said to you was between you and us

G repeating/clarifying that the interview content was not shared

Is he warning me not to bring in to this discussion what I picked up from the separate meetings?

Me Yep

P8M but we didn't discuss at all

G again emphasises the point

P8F No, no, we didn't, but that is us, you particularly, because that is what happened when I did the counseling, you were never, you didn't go to counselling but you don't have to use it. G said to me that he would come with me and support me but he knew I had every right to do that because I was worried about the children. And by then a certain amount of it I was worried for myself.

J agrees suggesting that is typical of them (not discussing together?) and links this behaviour with how G did not discuss the counselling she had pre the genetic testing. J respects his right not to discuss it together and makes point that she had to get tested because of her childrens' interests and she also acknowledges that she needed individual help for herself

Me When are we talking about?

Me wanting to be absolutely clear what counselling we are talking about and when

P8F This is when I in 1996 when went up to X before I was diagnosed and he came to every counselling session. We didn't discuss the counseling at all and I didn't realise until the day we got the results that he had not used the counselling at all because the shock I will never forget that. The shock on his face when I tested positive, it was absolutely amazing. And in fact he never came to counselling again did you?

J clarifies it was the pretesting counselling and says G came to sessions but they didn't discuss the sessions together as though he hadn't taken in the full import of the messages given because he was so shocked when the + result came through. In fact J implies that this experience put him off counselling again

P8M No, no

G agrees he did not involve himself

P8F Never ever. I went up twice afterwards on my own and H (daughter) came with me, she didn't come in but she came to (hospital) with me

J says 'never, never' with feeling and says how her daughter H came to (hospital) with her but didn't take part in any sessions

H presumably had her own thoughts about testing to cope with

The second stage of analysis involved extracting the codes and listing them on sheets of Flip Chart. This enabled the codes from each set of interview transcripts to be compared with one another. From this second level of analysis three overarching themes and their sub themes were identified.. This analysis was supported by detailed review of the process in supervision.

3.5.13 Reflexivity

Diary notes were made before and after interviews. These summarised my thoughts, reflections and observations of the consultation process and were later referred to in analysing interview content (Hammersley and Atkinson, 1983; Smith, 2008).

3.5.14 Trustworthiness and rigour

In pursuing and analysing the fieldwork for this study an attempt has been made to adhere to the recommendations of Yardley who gives four general criteria by which results should be assessed (Yardley 2000). These are:-

a) that the study should demonstrate sensitivity to context. b) that the results should be judged against interview quality and completeness of the analysis c) the study report should show transparency and coherence, and d) that the study should demonstrate a contribution to the field.

Chapter 4. Results

4.1 The data set

The experience of seven couples was explored in this study, four of whom were seen together in different combinations. Ann and John have already been referenced as one of the two couples consulted for the Scoping Review. They were interviewed once in a joint interview. Two couples (Peter and Kelly/ Jane and George) completed the full set of separate individual and a joint interviews. Sarah was seen individually but arranged for me to interview her husband in residential care for a brief twenty minute discussion.

Rachael, Helen and Henry were interviewed on their own. The partners of Helen and Henry had both died within the previous three years while Rachael was clear that Sam, her husband, would object to being interviewed and this resulted in being the one interview not to have taken place in a family home. The participants, how they engaged in the interview process and some family/medical details are summarised in the table below.

<u>Participants</u>	<u>Interviews</u>		<u>Length of partnership</u>	<u>Date of Diagnosis</u>	<u>Time since diagnosis</u>	<u>At Risk Children/ grandchildren</u>
	<u>Individual</u>	<u>Joint</u>				
<u>Couple 1</u> John (72) Ann (65) *		Dec 2010 Scoping Review	34 years	2004	6 years	Ann-2 boys 5-gchildren
<u>Couple 2</u> Peter (68) * Kelly (60)	One each Feb 2014	Joint Feb 2014	34 years	2007	7 years	Two girls 26/31
<u>Couple 3</u> George (66) Jane (70) *	One each September 2015	Joint October 2015	31 years	1996	19years	Jane-3boys 47/48/39 5-gchildren

<u>Couple 4</u> Sarah (51) David (55) *	One each October 2015		33 years	2008	7 years	Two girls 13/19
<u>Couple 5</u> Rachael & Sam *	One September 2015		30 years	2014	20 months	3 boys 28/26/24
<u>Couple 6</u> Helen & Mark* (deceased)	One October 2015		30 years	2003 Husband died 2012	12 years	2 girls 25/22
<u>Couple 7</u> Henry (77) & Tess* (deceased)	One October 2015		37 years	1995 Wife died 2013	20 years	5 girls/1 boy

N.B. 1. Identities are disguised with pseudonyms. 2. The asterisk denotes the partner with HD. 3. The numbers of children and grandchildren refer only to those who might be at risk of HD and excludes any children non-HD partners may have had with other partners.

The sample of couple and individuals interviewed was diverse in many respects and they can be thought of as clustering in different ways.

4.1.1 Couples living together in their own homes.

Three couples (John and Ann , Peter and Kelly, George and Jane) and one individual (Rachael) were able to provide accounts of the day to day challenges of living with HD when currently living together with their partners at home. In this group of four couples, Rachael's situation stood out because she and her husband were at an earlier stage of managing the illness than the other three couples and her husband was finding it very difficult to accept that he had the disease and refused to engage with any activities which recognized his illness.

4.1.2 A couple living apart.

Sarah and, to a lesser extent, David provided accounts of how HD had impacted on their family life and of how they were now adjusting their partnership to accommodate David living separately in specialised residential care.

4.1.3 Bereaved partners.

Helen and Henry had both lost partners to HD within the previous 3 years. Their accounts of their couple and family histories need to be understood, in large part, from the perspective of grieving their loss.

4.2 Couple similarities and differences: the stage of illness and some implications for accessing couple experience.

The foregoing clustering of the participants illustrates how participant couples are located on different points of the illness path. Rachael and Sam (C5) were in the early stages where Sam, for the most part, was denying that he had an illness and refused to be interviewed, resulting in Rachael and her adult sons carrying the main burden of care. Their position contrasted with those of Helen (C6) and Henry (C7) who had both lost their partners to the disease within the previous three years and were coming to terms with their losses. Although at very different points of the illness trajectory, the experiences of these three couples meant that couple functioning was seen from the vantage point of one partner, contrasting with the other four couples where both partners were interviewed. While Rachael relied on recall of HD related events that were either current or took place in the previous five years, Helen and Henry recalled HD related events which went back decades. Moreover, in relying on memory in different ways it was apparent that different emotions informed their accounts. Rachael, for example, was explicit in referring to her high levels of anxiety in managing the consequences of HD while Helen and Henry's accounts were both imbued with sadness for the partners no longer in their lives.

In regard to four couples -John and Ann (C1), Peter and Kelly (C2), George and Jane (C3) and Sarah and David (C4) – it was possible to interview both partners. (For reasons discussed later, the interview with David has not been included in the data set because the route by which he came to be interviewed did not conform to the research protocol.) Sarah and David aside, the other three couples were at the time of interview, at stages of the illness where

symptoms were not changing rapidly and where it was possible to gain their views about past, present and future aspects of their lives together.

Although the participants in this study were different in many ways, they shared common characteristics which the table above conveys. The first is that the interviewees are all over 50 years of age and all of their partnerships are, or were, longstanding with the shortest being of 30 years. The second common characteristic is that all of the participants had at risk children and, in addition, two couples-John and Ann and George and Jane-made explicit their concern about at risk grandchildren. The partnerships in this sample are, or were, durable and resilient, notwithstanding the many stresses to which they are, or have been, subject.

4.3 Overarching and sub themes

4.2.1 Knowledge of the family history of HD

- Not knowing about the family history of HD prior to diagnosis
- Ignoring the family history of HD and going ahead with having children
- Knowing about the family history of HD prior to diagnosis

4.2.2 Talking or not talking about HD- between partners and within the nuclear and extended family.

- Partners not talking
- Partners talking
- Talking to children
- Talking with the extended family

4.2.3. Negotiating the transition from a pre-illness to an illness dominated relationship

- Balancing anxious concern with a respect for independence
- The effects of HD on couple intimacy; the roles of carer and being cared for
- The move to residential care

The discussion that follows takes each of these overarching and sub themes illustrating each with quotes from relevant transcriptions.

Overarching theme: Knowledge of the family history of HD

Sub theme 1: Not knowing about family history of HD prior to testing/diagnosis

Four of the couples talked about ‘not knowing’.

Rachael (C5):- *“It was an absolute nightmare.”*

Rachael’s words capture the experience of four of the couples where a lack of knowledge about any past family experience of HD compounded the problems of coping with the disease’s early onset; the father’s of Sam (C5) and Peter (C2) were both likely to have been HD positive but both died from other conditions before becoming symptomatic. David’s (C4) father spent much of his adult life in a psychiatric hospital and his underlying condition was only acknowledged in the family when, at the point of David getting diagnosed, his family checked his father’s death certificate where HD was given as the cause of death. Sarah, David’s wife, suspects that there was family collusion to avoid acknowledging the family history until it was made necessary to do so by David’s diagnosis. Henry (C7) had no clear idea of how HD had impacted on Tess’s family whose roots lay abroad.

Rachael and Sarah (C4) were able to be fairly precise for how long they had to contend with difficult consequences of HD before a diagnosis offered an explanation or confirmed their growing suspicions. In Rachael’s case she reported that from about five years before diagnosis Sam was having difficulties getting things done, suggesting that apathy was beginning to appear and, for three years before diagnosis, she described his personality beginning to change in troubling ways. At the point of seeing a neurologist, she reported:-

“I can’t deal with him, I can’t cope with him, he’s now started to push me, if I said something he would have grabbed hold of me, he even broke my necklace-he didn’t have me around the throat but he was trying to tell me I was talking a load of rubbish and I didn’t know what I was doing”. Rachael (C5)

Her neurologist ruled out Parkinson's disease and MND but, before his diagnosis was finally made, suggested that Sam might be experiencing a 'nervous breakdown'.

Sarah (C4) said that the unexplained disease impacted on her family for fifteen years before David was diagnosed six years earlier. During the period before diagnosis, his mood changes, principally involving impulsivity and aggression, were becoming increasingly problematic. He was unemployed for four years before his diagnosis was made and, even before that, his employment record was erratic so that the main burden for supporting the family financially fell to Sarah. In this period of uncertainty about what was happening, Sarah had various tentative explanations. She wondered whether he might be seriously depressed but his GP ruled it out and was generally unable to offer a clear diagnosis or signpost for getting one. The internet became an important resource both to help her gain advice about how to manage his violence and also to form an idea of his illness by checking his symptoms against various website listings.

Rachael was not alone in considering a range of possible alternative diagnoses to HD when there was no family history. Henry and Tess (C7) had recourse to an acute psychiatric in-patient unit for several years before a particularly violent attack by Tess on Henry prompted doctors to make a referral to a neurological service. Until that point Tess was treated for what sounded like acute paranoid delusions coupled with what Henry called photophobia; it appears that Tess collapsed or had fits when exposed to flashing lights. Whether or not all Tess's symptoms could be explained by HD or were the result of other parallel conditions cannot be established retrospectively. It can be said that her positive diagnosis for HD influenced her subsequent pharmacological treatment and had important consequences for her six children who now had to contemplate the implications for their own health.

As was the experience for Rachael, making sense of unpredictable, often angry and violent behaviour was a challenge for other non affected partners and made more difficult by not having a diagnosis. While hearing a diagnosis was often referred to as having a devastating impact- (Jane (C3)-*"the shock I will never forget that. The shock on his face when I tested positive, it was absolutely amazing."*) -there could be benefits. Kelly (C2) described her two daughter's relief at now having an explanation for husband Peter's very angry out bursts which had been a source of great embarrassment to them, particularly in social situations,

during their adolescence. Memories of Peter's failures to turn up and provide a family taxi service for their girls and of angry outbursts in restaurants now became more understandable and forgivable.

Sub theme 2. Ignoring family history of HD and going ahead with having children

Two couples acknowledged that they had been told early on in their married lives about the risk of HD in their families but had gone ahead with having children. This seemed a particular problem for these couples because they said that, had they known then what they know now about the nature of HD and its heritability, they might have taken different decisions about having children in the conventional way.

Helen (C6):-

"...we all went along to a particular appointment and they- this is before his mum died- and they said that they thought it might be HD, the implications were that it could be passed on to children. But because I think it was kind of, we also went into denial...We just didn't want that and so we just sort of went ahead and had two children anyway. Also makes you think, perhaps that wasn't such a good idea. I think there were few options at that time".

Jane (C3):-

"...Grandad's doctor had said at one time when my uncle, my mum's brother was up there, looks as though he is going on the same way as his dad, and that did, did sort of put warning bells out...my (first) husband and I weren't engaged then, we weren't married, so it didn't ring any bells to not have a family until after I had had (my youngest). I said to (my husband), I think possibly we are chancing our arm with this Huntingtons thing because we didn't think Mum had got it because she is fine."

These two quotations from Helen and Jane's interviews offer helpful insights into how it was possible to go ahead and have children half knowing that, in doing so, they were running the risk of passing on HD to their children.

It is important to recognise that at the time their decisions were made to have children the state of public and medical knowledge about HD was far less developed than it is now. Jane's family doctor, for example, questioned why she wished to be sterilised after having her third child, claiming that HD was only passed on through the male line.

Jane (C3):-

"So I thought, now, do I just tell him or do I get angry, then I just told him, and I said no, no, no-it is on chromosome 4. I said, women are just as likely to get it as anybody else, 50/50%."

Moreover, at the time Jane and Helen started families there were not the diagnostic and counselling services that there are now; Jane's youngest child was born in 1976 and Helen's in 1993 and it was not until 1993 that a test for HD with back up counselling was introduced. In addition to a lack of information and testing services, there were, as Helen says *"few (alternative) options at the time."* Helen and Mark were not in a position to consider the current option of pre-implantation genetic screening (PGD) as a part of in vitro fertilisation treatment; a procedure that allows for only HD free fertilised embryos to be implanted in the mother's womb thus ensuring that any subsequent children are HD free (Human Fertilisation and Embryology Authority, 2017).

Moreover, had Jane decided against having her own children, it was likely that she would have felt under pressure to give an explanation to her mother, who held the view that HD was only passed down through her brother's side of the family. This challenge to her mother's opinions and beliefs was not something that Jane felt she could pursue and is one example of how a couple's management of HD threats to family building could affect relationships within the wider family and, conversely, how family assumptions within the wider family could shape the way couples approached HD risk.

Sub theme 3. Knowing about family history prior to testing/diagnosis

One couple-Ann and John (C1)-were fully aware of the family history of HD before Ann was tested. Her approach to being tested was strongly motivated by the hope that it would prove negative and that she would be in a position to reassure her sons by her first marriage that they were not at risk.

John (C1):-

“ The main reason Ann had the test was because we knew her father had it and so she could say to her son, that he didn’t have it-couldn’t possibly have it-because Ann wasn’t a carrier-but, unfortunately, I knew for months she was a carrier because of her facial expressions, whatever.. ”.

Ann and John knew about the risk of HD for several years before Ann was tested positive. Her father had developed symptoms fifteen years and died ten years before our interview. Gradually Ann developed symptoms, one of which-facial grimaces-convinced John that she had the disease although Ann approached the prospect of testing with the strong hope that she would prove HD negative and, thus, be in a position to reassure her eldest son that he could not develop the condition. The positive finding was, according to John:-

“a devastating revelation to you.”

John’s comment was confirmed by the subsequent comments in the interview which included many references by Ann to her eldest son who had by then begun to show signs of developing the disease and was described as having gone through a great deal of personal turmoil including a divorce and problems with alcohol.

Ann and John’s experiences were interesting in two ways. First, the knowledge of Ann’s father’s having HD helped John anticipate and be prepared for Ann’s positive test result. This was not the case for Ann who, in spite of developing symptoms and with the knowledge about her father’s diagnosis, held on to the belief or hope that she would test negative. This wished for result and its connection with the welfare of her son, explains the devastated reaction when the test proved positive.

Overarching theme: Talking or not talking about HD- between partners and within the nuclear and extended family.

Once a diagnosis is made how that information is dealt with at individual, couple and family levels emerged as a recurring theme for the participants in this study.

Sub theme 1. Partners not talking.

Five couples conveyed in different ways the problems they had in talking about HD and related matters.

Jane (C3), referring to how she and George didn't discuss her pre-test counselling:-

"No, no, we didn't, but that is us, you particularly, because that is what happened when I did the counselling, you were never...this is when, I, in 1996 went up to hospital before I was diagnosed and he came to every counselling session...(but) ...we didn't discuss the counselling at all and I didn't realise until the day we got the results that he had not used the counselling at all..."

Henry (C7) referring to his deceased partner, Tess:-

"You wouldn't have known really she had it until the day she died. She wasn't that communicative, she hadn't been social to talk about these things. Couldn't really approach it to be honest."

Sarah (C4) referring to her husband, David:-

"He would make suicidal threats and he is a keen motorcyclist and would hare off at very high speed along the road I described to you...or take off in a fury...it was bizarre, and I couldn't make sense of it, couldn't get him to see a doctor...If I challenged him.... that is when I got a black eye."

Rachael (C5) referring to her husband, Sam:-

"If I go back at him again he will really be difficult and if I go back again and again that is when he will actually take hold of me. I don't do that anymore...so we went behind his back, I feel really bad, I feel I am deceiving him, she said (referring to an HD advisor) you are not deceiving your husband you are going behind the back of HD, you must always think that, you are going behind HD's back not Sam's back. And we went ahead and did it."

Helen (C6) referring to her deceased husband partner, Mark:-

“He never ever thought he had it (HD)...even when he had to go into a nursing home. So that was my big problem in that Mark never ever noticed he had any symptoms. He never accepted the fact.”

For Jane (C3) and Henry (C7) there was a suggestion that the difficulty in talking about HD was a typical feature of their respective partnerships. Jane’s comment *“but that is us”* draws attention to an avoidant tendency between her and George that emerged at the point of anticipating the result of her HD test. As George said in relation to the build up to the test:-

“...at the moment there are no symptoms showing, we will plan for it. We never did plan for it and it just came, it hit.”

Henry’s (C7) comment that *“She wasn’t that communicative”* suggested that Tess had always had a communication problem which made HD impossible to confront.

There was no evidence of undue communication problems in Helen, Sarah and Rachael’s partnerships prior to HD. All the evidence from the interviews indicated strong and satisfying partnerships where the capacity to share joint concerns and interests stood out. However, in all three partnerships the male partner found the diagnosis of HD very difficult to accept. Denial was the defence that all three men used to varying degrees and, in its most rigid form was expressed by Helen:-

“He never accepted the fact.”

Sarah’s (C4) comments refer to the process of denial existing long before a diagnosis was made. Problems arose in getting David to see a doctor and, if a confrontation was risked, uncontained emotions were released. David’s threats of suicide linked to frantic and anxiety provoking escapades alternated with his violence against Sarah if he and she stood their ground. Finding a way round this flight/fight situation was a challenge for all three couples. One solution was described by Rachael who, with the support of her sons and the Regional Care Advisor of the HDA, *“went behind his back”* helped by the justification that in doing so

she was not relating to Sam as a person but to HD as an entity that itself had agency and needed to be dealt with; she was going “*behind HD’s back*”.

Sub theme 2. Partners talking

The two couples, Peter and Kelly (C2) and John and Ann (C1), who demonstrated a capacity to talk together about the impact of HD on their relationship also happened to be active and involved members of their local HDA branch. Whether their involvement facilitated their capacity to talk openly together or whether their resilience in talking openly made them the sort of people who would engage with an HD support group is an open question.

Kelly (C2):-

“It was devastating there is no two ways about it. I just felt so bad there wasn’t anybody to support, I felt so bad that poor Peter had to come out to the car park and drive himself back knowing, knowing that...I just wished I’d been there (at the appointment where he was told his diagnosis of HD)”

Peter (C2):-

“I suppose my greatest feeling was tears and I didn’t know what support groups at that time, but soon got them but my greatest concern was passing the gene on ...So nice to go out socially with a couple for a meal or something, again you don’t explain yourselves and they can say things like you ought to try counselling, things like that, it is of mutual benefit, a good friendship.”

Ann (C1):-

“I felt devastated, Chris, I know my life expectancy is not good...I just live for every day, I look after him.. we have a dishwasher..”.

These two couples conveyed an ability in their interviews and outside them to confront HD together and to find ways of talking about the impacts it had on their relationship from the time when the diagnosis was made. Although Kelly was unable to be present when Peter was

diagnosed at hospital, they quickly became involved in HDA support groups, partly to think about the implications for themselves but also to help prepare talking with their two daughters then in their early adult years. This openness to thinking about the disease's impacts has been reinforced by forming a friendship with another couple where the wife and Peter are at similar stages of the illness.

Ann and John were also able to convey in our joint interview a capacity to talk about the impact of HD on their relationship. Ann's feistiness to assert her independence alongside her ability to acknowledge her vulnerability, as illustrated in the quote above, coupled with their joint assertions that they have a good supportive relationship with family and friends within the local branch of HDA, all suggested that they were able to remain open to talking together about how HD impacts on their lives.

Sub theme 3: Talking with children.

Six couples in the sample volunteered information about how they had talked or not talked to their children about HD.

One of the dilemmas of parenting is knowing whether and how to introduce children to painful and difficult news that will have a profound effect on their lives. Resolving this dilemma involves balancing the need to protect children from knowledge that might be difficult to understand or tolerate, alongside recognition that, as children develop and mature, they have a right and a need to be told certain things that make sense of what is happening in their lives. One of the factors that influence the resolution of this dilemma is the age of children. All of the participants in this study had children who were likely to be at risk of developing HD and, at the point that a diagnosis was made, their ages (from those that were recorded) spanned from the youngest at six years to the eldest at twenty six.

At the younger end of the age range, Sarah and David (C4), Helen and Mark (C6), Peter and Kelly (C2) had six children (all daughters) between them at home at the time diagnoses were made. Sarah's daughter at age six was the youngest and Kelly's daughter at nineteen was the eldest. All of these six children had been exposed to and influenced by their father's deteriorating health, typically involving large mood swings and unpredictable/impulsive

behaviour. The following quotes illustrate key themes in this exposure, what and how they were explained.

Sarah (C4):-

“If (he) doesn’t consent, that blocks the family for a while...he didn’t want to tell the children, wanted to protect them...I eventually persuaded him to tell the children as they asked questions, and the younger one funnily enough is the one that wants to know everything. The older one firmly put her head in the sand. And I think did come to terms with it when she realised that there was a reason for Dad’s anger and behaviour, because they were impacted by his behaviour as well...they witnessed some difficult stuff going on with me. I mean I had a black eye, I had bruises on different occasions...he just didn’t know his own strength.”

Helen (C6):-

“I have never said anything to them about it...(and later in the interview)...Mark, if you don’t get out the car, then I am going to go over there and I am going to tell the girls what has happened because I have never said anything to them about it. So I am going to tell the girls why you shouldn’t be driving. I am going to count to two, if you don’t get out the car that is what I am going to do. Any rate, he wouldn’t get out the car so I called the girls over and I just said your Dad shouldn’t, the reason we are having this argument, I said, is because your Dad shouldn’t be driving. Because the doctor says he has got problems with coordination, I never told them what it was..... Anyway, with that he got out the car, slammed the door and just got in the back seat, so we all drove home. So that was the first time the girls had been told there was anything wrong with him. That was a difficult couple of days”

Peter (C2):-

“My greatest concern was passing the gene on and...(following the diagnosis) and telling the girls.”

Kelly (C2):-

“Now that (the diagnosis) explains things and as it progresses, it (HD) is very slow, as you know, we did, certainly myself and the girls have said, now we look back, we can see where, you know, this anger is coming from, it goes back quite a bit...Interestingly our youngest daughter (then nineteen) wasn’t that bothered by itshe said because it is not cancer... when you said I’ve got this bad news, she said my first thought was that it’s some terminal illness but while I know it is bad she said at least you are here and, you know, we can work through it.”

These extracts capture some of the complexity involved in talking to children about HD. Given David’s (C4) reluctance to accept his diagnosis it was surprising, and perhaps admirable, to hear that he had been able to talk directly to his daughters about his health problems. The responses of his elder and Peter’s younger daughter underline how making sense of what is told takes time-the initial “*head in the sand*” approach of David’s daughter gradually giving way to a realisation that his diagnosis made sense of his violent treatment of her mother. Peter’s younger daughter expressed an optimism which may have been motivated as much by a wish to reassure her parents as herself and she emphasised that a process over time was involved:-

“we can work through it”.

Helen’s account conveys a sense that she had misgivings in naming Mark’s condition and it required a crisis around driving their family car to produce the impetus to give her daughters a partial explanation of his problems. If Helen’s reticence was right for her girls at that stage in their lives, Kelly implies that the diagnosis helped her and her much older daughters understand their father’s difficult behaviour. Their daughters at age nineteen and twenty four were entering adulthood at the point HD was confronted and both Peter and Kelly were quickly aware of the implications this knowledge had for the girls needing to take responsibility for their own health and for current and future relationships. In helping them do this, they were candid in talking with them.

In reviewing the transcripts of those couples whose children were well into adulthood when diagnoses were made, it is striking that there were understandable pre-occupations about the possibilities of the disease re-emerging in the next generation and concern that, where possible, family planning was considered in the light of inherited risk. Jane’s three and Ann’s

two children by their first marriages, Rachel and Sam's three children and Tess's six children by her first marriage were all of adult age when HD had to be considered as a risk factor in their lives. In the extracts from interviews that follow, it is apparent that their children had mixed views about going forward and being tested for HD.

Jane (C3):-

"I didn't want my children to go ahead and have a family without some knowledge there, that was quite true, but I did also want to know what my future was, and, yes, we both cried over that, and the worse thing was telling the children really."

Soon after receiving the shock news of being HD positive, yet at a point she was symptom free, Jane and George made contact with Jane's three children to explain the implications for them. Jane is consoled by the thought that her own symptoms have emerged late in life and hopes that her eldest child, a son, who has tested positive, will have a similar late onset of symptoms. With his wife, he has used pre-implantation genetic screening to build a family. Her other two children, one of whom has a son, have not been tested, preferring to live with the uncertainty as to whether they carry the HD mutation. The various levels of uncertainty that this produces reinforces the need for Jane to be in regular contact with her children so that they can all monitor each other's health alongside sharing other family news.

Ann, to wrestled with the dilemmas about talking with her sons.

Ann (C1):-

".... the eldest (of two sons he tested positive for HD) is suffering at the moment... He has got a new girlfriend"

Me- *"Did the otherget tested?"*

Ann- *"He and (his partner) talked about it...he doesn't want to (get tested).....he works on a building site and says someone could drop a lump of concrete on me...."*

John- *"his kiddies might be affected-there are 3 daughters...They all find it difficult....we have always been very close and have a supportive family."*

In this passage above, John and Ann both refer to their “*supportive family*” implying that there is a full awareness of HD as an issue that is talked about and managed among those at risk. Both sons have made different decisions about being tested with the younger one deciding against it and preferring to live with uncertainty.

Rachael talked about her son’s attitudes towards the disease.

Rachael (C5):-

“He (her eldest son) can't deal with it mentally-he can't talk about it... he said I know I am going to die some time, which is exactly the same as what Sam says, I am going to die some time. (He) will be tested I think because he said to be fair to (his female partner)...I can see signs that (he) could possibly have it. None of them has been tested... all three are convinced they have got it. They have convinced themselves.”

Rachael’s family were, compared to other families in the sample, at a relatively early stage in coming to terms with HD, the diagnosis being given less than two years before the interview. All three sons were fully aware of the implications for them, none of them had children and, according to Rachael, all of them were against getting tested imminently but were anxious about getting the disease. Her concerns focused particularly on her eldest son who, she thought, was beginning to show symptoms but so far had rejected the idea of having a test. However he had said he would envisage doing so at some later date in order to satisfy his partner’s concerns.

Sub theme 4. Talking with the wider family

A diagnosis of HD can have important implications for relatives beyond the limits of the nuclear family and it was interesting that four couples elaborated on the different ways this information was handled. First, it was decided that some family members should not be told about the diagnosis.

Jane (C3):-

“And also, you see, because my mother didn’t know her status and was never going to, we had to talk that through in the counselling quite carefully because she was just in her 80s when I was tested and she was convinced obviously because of the fact that she appeared to be fine, but she was concerned, would have been concerned, about the grand-children and there was absolutely no point in letting her find out in any way shape or form. So we didn’t.”

George, Jane’s husband, reinforced the point:-

“And also it would have been a breach of confidentiality because, if you had told her you proved positive, it automatically would have meant that she is positive...So you are disclosing information that she has not got a right to know...as far as she was concerned she was not symptomatic.”

Keely and Peter (C2) held similar views:-

Kelly- *“Fortunately because of the relationship with my mother-in-law, it wasn’t something, I think, I don’t know whether she suspected that there was anything wrong as such, I think she thought it was severe stress as well, but if she had been of that type that, if we had got together over it, maybe we might have said-should we ask him to go to the doctor-or something, but because I wouldn’t have been able to have said anything to her about it, having said that, but it’s a relief she never lived to know about HD because it would have destroyed her.*

Peter- *“It would have destroyed her world.”*

It was agreed by these two couples that Jane’s mother and Peter’s mother should not be told about their childrens’ diagnoses. In both families it was believed that to have given this information would have been devastating for them. The decision to keep the diagnoses secret required the cooperation of other family members, a position which contrasted with both couples’ determination to be as open as they could be with their children.

Second, a diagnosis of HD in one part of a family could require a degree of open communication with the extended family for a number of reasons. Rachael felt a strong obligation to spell out the genetic risks with parts of the extended family which share a common HD blood line. In this she was encouraged by their neurologist.

Rachael (C5):-

“(The consultant neurologist) said we must tell the family they are at risk and Sam took that on board really well... he is one of seven children, he has got three brothers, two sisters, one sister has actually died, and they are in complete denial, as well, of the illness. They notice no symptoms, no signs...but they are in denial now and I can feel there is beginning to be a rift between me and the family...he actually phoned his brother, went to meet his brother and he told his other brother. We didn't tell his mum, his mum is 82. We didn't feel she could cope with it (but) yes his mum came up and she said to me, is it anything serious? She said it is killing me not knowing and she started crying which is me, her and Sam's sister. And brothers were in the other room and I just went in I said it is not fair on her, I am a mum, I have got three sons and it is killing her anyway, she is hurt anyway, so do you think we should tell her? So we backed Sam into a corner, all of us, and we told him he should, we felt it should come from him but if he didn't want to tell her, then one of us would, but we felt she should know now. And he told her, she doesn't understand it all. Whether it was the right decision or not I... she does know now anyway but she does know, yes, a hard one really.”

One of the consequences of Rachael falling out with parts of the extended family was that it undermined the possibility of them offering support as Sam's health deteriorates in the future. Estrangement from the extended family was also Sarah's (C4) experience. Her wish to be able to talk about HD was prompted, less by a need to alert mother-in-law and sister-in-law to the health risks of HD (David's sister had tested negative for HD and it had been his father rather than his mother who carried the disease) but from a wish to cooperate over David's care. This was proving impossible as the following quote demonstrates.

Sarah:-

“His family still aren't talking to me at all. I had one or two really angry episodes...I think they can't visit it...can't handle it...his mother has been through the same thing and my

assumption is that his mother had been asked to separate from his father...and, she has never faced up to it. She buried her head in the sand. And I have sympathies with his Mum but I haven't had a lot of sympathy from his sister who I understand has had survivor guilt...and I would try to give them a book. I remember giving his aunt a book about HD because the poems that the HDA sell, but the comments that I got back was I can't read. What do you mean, can't read? My vision is too bad. I knew her vision wasn't great but...what sort of excuse is that?"

Overarching theme: Negotiating the transition from a pre-illness to an illness dominated relationship

Sub theme 1. Balancing anxious concern with a respect for independence.

A progressive degenerative illness of whatever sort presents a massive challenge for couples. Independent activities for both can be restricted as the competencies of the ill partner diminish and an increasing burden of care is placed on the shoulders of the stronger partner. In the quotes taken from interviews with five of the participant couples, it is possible to discern the struggle each couple had in balancing care and concern, on the one hand, with a respect for autonomous independence, on the other. In most partnerships the care and concern was expressed by the well partner and the bids for autonomous independence were made by the HD partner.

John and Ann (C1) commented:-

John- *"Because of Ann's deterioration I had to retire last December...perhaps I am too protective."*

Ann- *"I bath myself every day...shower every night...I like being on my own".*

In John's words he questions whether, in responding to Ann's repeated mini-crises in the home, he has become too protective. In reply, Ann was keen to assert her capacity to look after herself and to enjoy her own company. Much of the interview was a variation on this theme with John illustrating his caring role and Ann either asserting her capacity to do things on her own or to contribute as an equal partner to common tasks, like cooking. In the way she

repeated these claims, she conveyed the impression that it was a struggle to maintain her viewpoint.

Rachael (C5) said that:-

“He’s always provided for us, always made sure we are OK, always put us first. Never ever would he put himself first, it was always we were OK and then, Sam has changed. But when he had his licence revoked, he thought he’s fine to drive...(the problem is) his physical jerkiness I would say, slightly, I think that is the main reason, what will happen if somebody pulled out in front of him, he wouldn’t be able to respond...we have got a great big dent in the front of his car which he did...he said, oh it was just like it was dark and the car was all misted up. But his spatial awareness, he knew I was going to go mad because I hid the car in the garage...So it has been over a year now unable to drive and... (he) still doesn’t accept it, still fighting to get his licence back..”

In describing the way that, now, she has to look out and protect Sam, Rachael makes the point that this represents a massive reversal of how she and Sam related as a couple before HD came into their lives. *“Sam has changed”* from the husband who looked after others to a man for whom she now has to care. Sam’s stubborn insistence on maintaining his independence by continuing to drive a car appeared unsafe to Rachael who is then forced to manipulate a situation to safeguard his wellbeing and that of other road users.

Helen (C6) recalling an earlier time said:-

“I would get phone calls at work saying from the police saying somebody has rung in because they are worried that there is a disabled man who delivered leaflets for you and he is walking down the road and...it is pouring with rain and he hasn’t got a raincoat on...so I would have to go and sort of like drop everything and go and pick him up and bring him back home.”

Helen respected Mark’s need to go out each day and distribute leaflets locally to advertise his services as an occasional gardener. This routine helped Mark maintain his identity as a

working man but it was also obvious to members of the public that he was sometimes vulnerable, requiring Helen to drop what she was doing and rescue him.

Sarah (C4) found herself having to rescue David on many occasions and also found it difficult to be independent of him:-

“I tried to support him through that although... I had a few little rescuing episodes as well. You know he would go out and have a puncture and phone me up to be rescued... My husband was very fearful when things were difficult that I would leave. There was a particularly difficult episode when I went to celebrate a friend’s 40th birthday... He was incandescent that I should leave him for four days with the children. I might get raped, I might go off with someone,, and it really was a very innocent girly, ok, slightly boozy, long weekend. But it really highlighted, looking back, his insecurities...”

While David had little anxiety himself in going away from home and getting into scrapes from which Sarah rescued him, it was more of a problem for Sarah to claim ‘me time’ as her description of events around her weeked-away trip reveal.

The need for ‘me-time’ was also referred to by George (C3):-

George- *“I have already seen two people stumble on those humps.”*

Jane- *“I have not actually fallen for absolutely ages.”*

George- *“Thank you very much, I have switched off now, go down the pub and don’t talk about HD.”*

While the struggle for independence was most frequently expressed by the person with HD and the experience of anxious concern lodged in the well partner, the process could also be reversed. George’s two quotes above express both his anxious concern about Jane falling over-something she is at pains to reassure him about-but also his need for getting away and having what he referred to as “*me time*” in the pub where he no longer thought about HD. In the joint interview he referred several times to his justification for claiming “*me time*” suggesting that his time away from home also stimulated a degree of guilt.

The interviews with George and Jane revealed that both have on-going chronic health problems resulting in them each having to be concerned for the other at various times. George had both a TIA and a perforated stomach ulcer in the recent past which meant that, in Jane's words:-

"We are a bit like a pair of old bookends holding one another up... you see... it would be just nice, sometimes, to be husband and wife and not carer and cared for. That's because the boots on either foot at the moment, you know,"

And the precariousness of this situation was made clear by George:-

"We are both only children, so Jane lost her father many, many years ago...I lost my parents in the 90s and the 00s...So we had no one else to share that burden other than each other, we have no siblings at all."

Sub theme 2. The effects of HD on couple intimacy: the roles of carer and being cared for.

Most of the participants reported adverse consequences for their sexual relationships and the mixed feelings that then follow.

In my joint interview with Peter and Kelly (C2) the following exchange took place:-

Me- *"... and for you Peter, how would you answer that question about the impact on your relationship together since the diagnosis?"*

Peter- *"Very sad, the move from being lovers to being cared for and carer."*

Kelly- *"And interestingly when I was having difficulties with our relationship somebody did say to me that if you try to think of yourself as a carer rather than a wife, you might find it easier, not to get so hurt, it is very easy to get hurt and I have to say, hard as that has been or was, it has worked, it has, it doesn't mean I am not loving, it does mean I am a little bit more detached mentally, so I don't get so hurt. They don't realise that they are saying things or not being attentive, that is a characteristic of HD, that you can be slogging your guts out and you*

can be sitting there, not aware that you are, which I know it is not you, obviously, it is the disease and I do find that the hardest thing now, not that I don't remember the good times, I don't know how to describe it, it is hard to remember those times, sometimes, when it was different, because it is going on so long, it is sometimes hard to remember, it wasn't always like this, obviously, but it can sometimes be a bit hard."

Me- *"And your comment in changing from lovers to carers, what sort of feeling does that engender in you, Peter, when you were telling me that?"*

Peter- *"It makes me feel a bit like second class."*

Rachael (C5), too, commented upon the loss of intimacy:-

"You lose your best friend, your husband everything...(in relation to their sexual relationship) but now it is non-existent. That has been about three months but it sounds awful doesn't it, but I am quite happy with not doing it and I think it is because it is the whole thing. But, yes, now I am thinking, but he, I mean, since we found out he is on a high calorie diet now, I have put on three stone. And he will say to me, well, I wouldn't want to do anything with you because you are so fat. I know I shouldn't be eating the high calorie food but I am a bit stressed. That is my escape now."

Helen (C6) also commented upon the loss of intimacy:-

(Mark started soiling which affected their intimate life) "We just kind of carried on doing what we had always done and everything was ok and then it got to the stage where when he started, basically, when he started... we got to the stage when I didn't want to have sex with him anymore and we didn't do it very often anyway by this stage. I was really worried that he was, it was, I didn't want to say no because I felt it was obviously he wanted to do it, but I didn't really want to do it any more so I kind of, and then one day I just thought, do you know what, I just can't do this anymore, and I just said, I really don't want to do that. And actually he accepted it and I was really surprised and I just thought, oh God, I have been worried about this for so long and I have been putting up with it for so long and I wish I had said it a year ago".

Sarah (C4) talked about the harassment she experienced:-

“He became quite aggressive late at night. I just wanted to sleep and hide and he pursued me physically. I just wanted to be left alone and that was really distressing for on and off for two or three years probably... I wanted to have some extra living space here so that the girls could have peace and quiet while dad was sat in the living room.”

Jane (C3) reflected upon her sense of loss at no longer having their former closeness:-

“It would be just nice, sometimes, to be husband and wife and not carer and cared for.”

The five couples from whose interviews the above quotes are drawn conveyed the distress and sadness that surrounded the relinquishment of an active sex life with their partners or an intimacy that was part and parcel of being husband and wife. Rachael, Jane, Peter and Kelly both speak about the sense of loss that accompanies this change with Peter poignantly expressing how this loss affects how he now thinks of himself as second class. It is noticeable that Kelly, Rachael, Helen and Sarah, all of whom had male partners with HD, talk about the relief that came from having ended an intimate life. Before this point was reached, their accounts point to the different ways in which intimacy could be linked with feeling emotionally hurt (Kelly) and/or physically harassed/pressurised and obligated into having intercourse (Helen, Rachael and Sarah).

It is also important to recognise that Ann and John (C1) were keen to stress that they continued to value the sexual side of their relationship and that it co-existed with having to manage unloving feelings at times.

Ann:-

“ In bad moments, you don’t love me you hate me...he gets quite cross with me sometimes (but) we are still a close couple.”

John- *“ If Ann lets me down in public like last week when trying to catch a bus and Ann thumped me...poor old thing... you are a two to three year old kiddie at times, you act like a three year old....”*

Ann- *"We had a good marriage sexually"*

John- *"I'm still keen"*.

Sub theme 3. The move to residential care.

Three couples had to manage their partner moving into residential care and the quotes that follow are long, reflecting the protracted and complex processes that led to residential care being needed. Shorter notes would not capture the significance of process and time.

Helen (C6)-

"I had to have him Sectioned...the problem was that he started soiling himself. And he didn't notice and he wouldn't notice but you could smell it. And of course the girls..(it was) really upsetting for them... and I remember coming home from work and (daughter) came home and she sat out in the garden and she said I can't go in the house. And I said, it is ok, I will sort it out. And I just basically rang ...social services...and said I need, I need my husband taken into care, I can't cope with him anymore because my daughter can't cope...and (eldest daughter) had previously had had a suicide attempt and so I just said, my daughter has tried suicide before and I am not going to go through that again... it was either him or them. I had always tried to keep the family together... I wonder if sometimes whether I did that for too long...and also, I kind of also felt, because the girls obviously have a 50% chance of having it, you sort of think, well you don't want them to think that if anything happened to them there wouldn't be someone to love them... so he was sectioned and taken into care, so it all worked out ok."

Sarah (C4):-

"I suppose, when things started getting really difficult and I was particularly exhausted he became quite aggressive late at night. I just wanted to sleep and hide and he pursued me physically...that was really distressing for on and off for two or three years probably... but (daughter) was doing her A levels by this time and it was really distressing me that he couldn't see when his behaviour was affecting her...that is when it came to a head. There was one particular episode... and he was incandescent...so we actually hid in the car and he was banging on the car with his fists trying to get us to come out and sort problems out and he

was really very frightening at that stage and in front of the children...then I thought I have really got to do something about this, I can barely function...I took advice from a national website which was new, realised I could take him to Court myself without a lawyer because I couldn't find a lawyer...they drew up a statement...I had asked for help from the mental health team ...but on this particular occasion they felt there was nothing they could do...he wasn't sectionable and they didn't think he was... (I went) to Court to get an injunction, to have the papers served, came back he was sitting in that window there, and he left the house fairly calmly."

Henry (C7):-

(Talking of care in the later stages of her illness) "Anyway, she then couldn't walk properly, she was there (a community centre) about 6 years, she liked it because there were loads of people about. And she had to go in a wheelchair, we even had to carry her on to the bus. Socially it was brilliant. But then we had this flat about the same size as this, two wheelchairs and all sorts of things and just haven't got enough room. Got to move...bungalow...had her in there and she deteriorated a lot...the thing about the bungalow is I never went to anybody. They didn't even know I was on the radar... The thing is, I had a full time job looking after her...and the bad thing about this was then when we were put on the radar, I never went and seen a doctor and they virtually said, who are you? You shouldn't be doing this on your own. Well I am. This is what I do. I don't ask for help...change her and all the rest of it. And, of course, then they put me on to a day centre, you must go to the day centre. So I said, oh alright then, we will go to the day centre, don't forget she had photophobia, she couldn't travel in a vehicle...Anyway, so I had to push her to the day centre, no transport, it wasn't very far but, the thing is, she was always frightened, the transport was there but I just couldn't use it...and then, of course, we had the extension built on the bungalow made even more room, and then, of course, we had to convert one of the bedrooms...it was two bed but one of the bedrooms was a traffic area, we couldn't use it as a bedroom so I used to sleep on the lounge floor. But the thing is it was important because I could hear her then in the hospital bed because I might have to get up in the middle of the night...well, she went into a nursing home. I wouldn't let her go and they kept saying she needed to go. That is a bit rich."

As these quotes above convey Sarah, Helen and Henry had to face the difficulty of helping their partners enter residential care under very difficult circumstances. At the time of the

interviews, the partners of Helen and Henry had died while Sarah's husband, David, was well settled in a residential complex and Sarah, David and their two daughters were adjusting to these new circumstances.

All three admissions to residential care were distressing. Two were the outcome of acute crises that required the well partners, Sarah and Helen, to turn to the civil law to evict their partners from the family homes. In these desperate circumstances, the recourse to law was triggered by crises in which both Sarah and Helen had to place the welfare of their children before that of their husbands. Sarah was forced into using adversarial legislation through the Family Court while Mark was assessed as sufficiently disturbed to be sectioned under the Mental Health Act.

If Sarah and Helen were placed in the position of having to evict their partners from the family homes, Henry had an equally painful account of seeking to hold on and care for his partner at home against the advice of professionals that she should be admitted to a care home in the final stages of her illness. Henry was quite explicit in stating that he thought Tess was taken from him by an uncaring and incompetent social care system. His account of their life together suggests that in spite of the difficulties presented by Tess being ill, Henry became highly skilled and fulfilled in caring for her.

Commenting on the time after their partners were required to leave home, Sarah (C4) said:-

"He went to his mum's. The family was shocked, horrified, said how could I do this. Could he stay with them while she studied? No he couldn't, my mum is 84, this is what sister says, and I am off work with stress. .. we were in crisis, Courts weren't very helpful because he had mental capacity and he could choose where he lived. They wouldn't get in touch with me because I was at odds with him in Court...I had to telephone four care homes...(having found one) he was there temporarily for 11 months because there was nowhere else suitable. Social workers would barely talk to me. I tried to continue to have a bit of a relationship to him as it were holding his hand no one else was his advocate...we fell through every gap in the system. He lost the ability to handle money...he lost that ability, he lost the ability to self care, shower and bath. So I was going in helping him shower and bath and he started to smell. Not nice to be next to...It was terrible then he physically went downhill, wet his bed. Then started wandering, then he started walking back here and he would bang on the door late at night or

early morning... It wasn't a place that was secure. There was one night he slept in our shed, we had been told not to let him in...and then a place becomes available where he is now, I was battling to find out about the funding, they wouldn't tell me and still he was wandering home. One half of me I am grieving him because I have lost my husband, I am still coming to terms with the fact..So I am gaining confidence in the staff there."

Helen (C6) remembered:-

"I went and looked at places so that I could recommend somewhere that I thought would suit him, because I didn't want him ending up in an old people's home... He did say to me,, when he went to this home, he said, I can't stay there, I got to come home. He said, I can't stay there, if I stay there, I am going to commit suicide. And I said, Mark, I said, that is completely up to you. I have tried as hard as I can and I can't do it anymore. You have stayed at home with me and the girls for as long as you physically could and we cannot have you at home any more, and so this is your life now. And this is where you live. I will come to see you, I used to go and see him in an evening once a week and then at weekends... I got so as I didn't really care anymore, and also I had realised that I could say things and he didn't actually get as cross as I thought he was going to get sometimes, especially by that stage. I had realised he was almost... it was basically just like talking to a child."

Mark died in the home.

"This might sound a bit of a strange way of putting it, but I think I was very lucky in the circumstances in which he died in that he died, I was just starting to get to think I am really going to have to do something about Power of Attorney...and then he died before it all happened so I kind of thought that is good, good of Mark. And also the circumstances were that, basically, he choked, but it happened in the day room so he was actually with people...and one of the nurses at the home, who I thought was very good would have known what to do..(Mark) didn't believe in euthanasia or anything like that. He always wanted to be alive for as long as he possibly could. And so I kind of thought it kind of like, he would have wanted, if anything could have been done, he would have wanted it to be done, and it was, but it didn't work. So it kind of worked out well for both of us...so I think that made it a lot easier to cope with as well. Yes, I kind of feel I have got my life back really. I kind of, when Mark went into the nursing home, it was a big relief. I didn't really feel guilty because I felt

like I did my best. And the thing that I found now is that I miss Mark now. Whereas when he first died it was like kind of a big release and it was all the things that had happened were very present, whereas now I miss the person who he was, you know... but he is not here. But I kind of miss the guy he was rather than the person he became."

Henry (C7) was very involved when Tess was admitted to a care home nearby:-

"Oh yeah, yeah, I visited her every day for two years. She died on about 7 December, two years after she... I had to take her into hospital she was choking, they were trying to, she had a peg by then."

(N.B. PEG is a form of feeding where a tube is inserted into the stomach through the abdominal wall thus bi-passing the mouth/throat and avoids the complications of choking)

Reflecting on the help he received from a district nurse and the carehome and comparing it to the care he gave Tess:-

"Well, help was causing me more trouble because, the thing I worry about was that, if I called somebody to assist me in an emergency on a regular basis they would take her off me. Because they would say, well he can't, he can't cope because if they didn't know. The one occasion when I did call for them because, of course, because she was being handled differently, it was the district nurse, she actually wet the bed and she had never ever wet the bed in all the time I have been looking after her...anyway we had our problems across there (the nursing home). She wasn't getting the right care that I was giving her at home. When she was put in a nursing home I had all sorts of problems, never stopped going to hospital, I have been ok. I think upset, they took her off me and didn't care for her properly. So can I show you some letters?"

A number of common themes stand out from these accounts.

It is clear that Sarah, Helen and Henry continued to love and care for their partners throughout the moves in to residential settings. In these demanding times, all three had to shoulder great responsibility. Sarah continues to feel angry that her needs were passed off by social work agencies so that she fell in the gap between them. Both she and Helen were

involved in selecting care homes that could accommodate younger male residents when most were targeting the needs of the very old. Henry felt that the social care services could not provide the quality of care that Tess needed and even felt that their care was negligent. When interviewed all three made references to the problems in coming to terms with the feelings of loss associated with these moves heightened for Helen and Henry by the deaths of their partners. Sarah said:-

“I am grieving him because I have lost my husband”

and Helen talked of missing:-

“the person who he was, you know... but he is not here. But I kind of miss the guy he was rather than the person he became.”

Henry talked of being:-

“upset, they took her off me and didn’t care for her properly. So can I show you some letters?”

Some of these letters were part of his continuing dispute with the care home about Tess’s care.

The offer to share with me written memorabilia of past times was also extended by Helen who had found some comfort in writing down what she called ‘Helen’s Rants’ during the period of approximately six years between Mark’s diagnosis and his admission to psychiatric hospital under section (See Appendix 4 for Ethics approval to use this data). The title captures the angry exasperation she felt at times but could not express because of a determined need to keep the peace for the children’s sakes. Responding to his complaints that she had disturbed his lie-in and accusing her of offering a half, rather than a full, cup of coffee she wrote:-

“Are you ill or not? If not then sort yourself out and stop trying to fuck me up or I’ll fuck off.”

But she did not leave him and stood by Mark as his behaviour and symptoms worsened. His insistence on driving a car when he was clearly a danger to himself and others, his failure to

tidy up in the home, his persistent sexual demands all took their toll on Helen who, at one point in 2004, wrote:-

“I just wish I was strong enough to find a way out of this but I’m afraid I can’t face it.”

It is difficult to know what solutions she might have been entertaining in her mind, but, whatever they were, her words convey her utter desperation and loneliness at that point.

Chapter 5. Discussion

Discussion of the results from this study will be in three sections. The first will take each of the overarching themes in turn and review how they and their sub-themes relate to the findings contained within the Literature Review. Where the results of the study do not have connections with the Literature Review other references will be introduced and their relevance discussed. The second section will be an appraisal of whether the aims of the study have been achieved and it will highlight the contribution this study makes to the current literature. The third section will outline the limitations of the study.

5.1 Results of the thematic analysis and their relationships to the findings of the Literature Review

5.1.1 Knowledge of the family history of HD.

Whether knowledge of the family history of HD was present before a diagnosis of HD was made and, if it was, how it was managed, emerged as an important overarching theme. In summary, three different patterns were identified. First, there were four couples who had no knowledge of the family history prior to a diagnosis being made. Second, there were two couples who had some lingering suspicion that they and their families might be at risk but had chosen to turn a blind eye to this knowledge influencing their lives. Third, there was one couple who had been aware of the family history of HD because a parent had died from the disease five years before interview but after many years of being symptomatic.

These three categories overlap with the findings of Etchegaray (2006) and Forest Keenan et al. (2013). Both describe different but overlapping trajectories of how individuals came to understand being at risk for HD. “Something is wrong”, (Etchegaray) and “Putting two and two together” (Forest Keenan) relate to that period when, in the absence of knowing the family history, there is a need to find an explanation of developing symptoms. When the result of this process was a positive HD diagnosis, there could be an experience of a shock coming “Out of the blue” (Etchegaray’s second category). Both authors also capture the experience of those individuals and couples who chose not to fully act on what they know about the family history of HD. “Knowing but dismissing” and “Turning a blind eye” are the

two phrases which Etchegaray and Forest Keenan use, respectively, to describe this process of conscious avoidance.

None of the participants in this study had the experience of “growing up with HD” which was one of the means by which some of Etchegaray’s research subjects learned of HD. This absence in the current study may be a reflection of the ages of participants all, of whom were over fifty years of age and grew up at a time when HD was not understood as well as it is today by professionals and the general public. Since the time of the participants’ childhoods, changes have come about on a number of fronts. The Huntington’s Disease Association was set up in 1971 and, through its publications, educational and support services throughout the UK, has helped de-stigmatise the illness and allow HD to be more openly talked about. The biochemistry of the disease is better understood. The identification of the defective gene was made in 1983 and the genetic test deriving from this knowledge was introduced in 1993 (Quarrell 2008). A further development of this work has been the possibility of breaking the intergenerational heritability of the disease by offering pre-implantation genetic screening (PGD) as a part of in vitro fertilisation treatment (HFEA 2017). All of these developments are relatively recent and, arguably, contribute to a culture where the sharing of knowledge about the disease becomes increasingly necessary to pass on to children so that they can make informed decisions about managing risk.

5.1.2 Talking or not talking about HD- between partners and within the nuclear and extended family.

While knowledge about the HD is readily available, whether and how it is accessed and processed within the nuclear and extended family emerged as an important over arching theme.

Partners not talking.

The inability of partners to talk about the illness together was a striking feature of the interaction of several couples at particular stages of the illness. The adverse neurological consequences of the disease cast light on why communication was problematic for four of the couples. The men of these couples behaved in difficult ways towards their partners; they could be irritable, angry, physically violent, sexually disinhibited, persistent in stubborn

denial of their condition and prone to apathy and depression. These symptoms are often first recorded in the early stages of the illness (Martinez Horta et al. 2016; Van Duijn et al. 2008) and, therefore, hamper the ability of partners to communicate freely in the pre-diagnosis stage. Complicating this picture was a difficulty some HD participants had in speaking and thinking clearly, a difficulty identified by Harletius et al (2010) who charted the way this language problem hinders communication in the family.

There was also evidence in this study that difficulties in talking about HD were partly shaped by psychological factors. Two couples talked about a lack of open communication between themselves, even in their pre-illness days, suggesting that this was somewhat typical of their interaction. Superimposed on characteristic ways of relating were the challenges of managing particular issues or situations that stirred up high anxiety. Typically these matters could be difficult to talk about in a reasonable way and might be managed, initially, by avoidance or denial. This strategy was often justified as being a way of preserving family cohesion in the interests of young children. However, this way of managing often broke down when the consequences of not facing a problem outweighed any benefits that might arise from ‘papering over the cracks’. The process of confronting problems was frequently very disruptive and painful, especially for children, although it was apparent for some families that learning about the diagnosis of HD could make sense of hitherto unexplained symptoms. However, situations and issues that stirred up high anxiety and were difficult to face included:- the anticipation of and coming to terms with the results of diagnosis and testing; facing up to the time when driving a car is no longer safe; confronting sexual problems; managing family finances; confronting issues around personal hygiene and contemplating residential care.

The identification of these specific problems flesh out the general finding of those studies that identify the mismatch between the ways carers and persons affected by dementia view their symptoms and their overall quality of life. There is a consistent finding across a number of studies looking at couple relationships, where one partner has either HD or another dementia, that carers, as compared to their partners, identify significantly more symptoms and feel that their partners overestimate the extent to which they are in control of their symptoms (Kapstein 2007; Ablitt 2009; Ready 2008; O’ Connor 2008). It is possible that this mismatch can be an outcome of communication difficulties (it represents a rationalization of underlying

problems) and/or is a contributory factor towards communication problems. Either way, carers report lower quality of life scores than do their partners.

Talking with children.

This study highlighted the very large and complex field subsumed under the heading of ‘talking with children’. Repeated references were made by participants to at risk children who, at the times when parents were diagnosed, ranged in age from six to twenty six. At the time the interviews were recorded, at least, three of the oldest in this group were in their forties and had established families of their own while the youngest in the group was studying for GCSEs. The rest were at various stages of independent living away from the family home. Given the significance of this subject it was striking that the Literature Review did not produce any references that focused on talking with children. This should not be taken to mean that there is no literature about the interests of children affected by HD. The HDA produces a helpful fact sheet on the subject and there is a lively website for the Huntington’s Disease Youth Organisation (see hydo.org) which encourages young people between the ages fourteen to twenty five to access information and to engage with sharing experiences. Given the broad range of childrens’ ages covered in this study and their very different circumstances and needs it is, perhaps, unhelpful to think of ‘talking with children’ as one undifferentiated challenge.

Talking with the wider family

The role of the wider or extended family in the management of HD is important in at least two ways. First, the importance of the contribution of the extended family in supporting couples and families contending with HD is well acknowledged in the research literature (McCabe and O’Connor, 2012) and there is some evidence that, in some families, family support actually increases following the emergence of HD (Aubeeluck et al. 2012).

However, the opposite can happen and there is evidence that HD can be associated with a lack of family cohesiveness (Vamos et al. 2007) which Domaradski (2015) lists as one of the negative impacts of HD within the wider family system. Second, the introduction of genetic screening of fertilized embryos as a part of IVF treatment in order to ensure HD free children has, according to Huniche (2011), generated a moral consensus or culture encouraging at risk young adults to engage with genetic information, services and technologies. She argues that

this culture places pressure to confront procreational risks before trying for children. This moral imperative is one of the key drivers that encourage some affected families to communicate with others within the extended family who, it is felt, need to be made aware of the genetic risks to themselves and any potential offspring. There was evidence in one interview of a neurologist encouraging this dialogue.

Within this study it was noticeable that this imperative to communicate was experienced in complex ways. There were examples of attempts to speak with members of the extended family which were rebuffed and/or rebutted as being based on false evidence. This miscommunication could be the result of a defence of denial within those parts of the family where good advice was being refused. The breakdown in communication had the further consequence that, as links with family were broken, the HD affected couple could feel isolated from potential sources of support.

However, the imperative to communicate knowledge about HD within the extended family could also be strongly resisted. Two couples in this study gave forceful arguments for why an elderly relative should not be informed of their child's positive test. In both cases it was argued that the elderly relative would not be able to come to terms with the news because in one case it would expose that relative to her own at risk status (a piece of medical information that she had not sought) and in both cases would generate devastating anxiety for those elderly persons in worrying about their at risk progeny. The experience of maintaining this family secret contrasted with the commitment to share the knowledge about HD with their children. From these examples it is clear that the challenges to communicate within families is both an intergenerational matter and a cross generational matter and that each of these dimensions contributes to the potential burden of responsibility carried by affected couples.

5.1.3 Negotiating the transition from a pre-illness to an illness dominated relationship

The Literature Review identified the fact that in the whole field of dementia studies a focus on couple interaction is in its infancy. Ablitt (2009) and Kitwood and Bredin (1992) make the point that for several reasons it has been difficult for researchers to address both partners as an interacting system. These include ethical concerns that research participation may cause

harm, there may be practical reasons for accessing affected persons and there may be problems for researchers to engage with behavior that is emotionally challenging, difficult or disturbing.

In this study it was possible to identify some elements of the relationship challenges that both partners had to confront and manage from the earliest stages of the illness through to managing the move to residential care, the death of a partner and coming to terms with bereavement. Three broad challenges were identified from the interviews

Balancing anxious concern with a respect for independence.

One of the consequences of HD is that families become more isolated as a result of social and employment connections being lost. Links with friends and family can become strained, work related friends fall by the wayside and opportunities for pursuing leisure activities diminish as family finances are squeezed (Domaradski 2015) (McCabe 2008). In addition to these significant losses, the limitations that HD imposes on the autonomy and independence of the affected person mean that the physical and emotional distance between partners closes. The HD partner is more often at home as a result of losing employment and is often joined by the non-affected partner who takes on a caring role. Under these circumstances the pre-illness balance point between separateness and togetherness is altered and it can be difficult for both partners to renegotiate a satisfactory physical and emotional distance from one another as both seek to find new ways of relating to each other and interests outside the family. This struggle has been identified in other contexts where a life changing event causes the emotional distance between couples to close. Mattinson (2008) discusses this phenomenon in relation to the impacts of unemployment on marriage.

In this study, the tension between separateness and togetherness was coloured by the particular challenges of a serious degenerative illness. It meant that, at any point, anxious concern (normally, but not always, located in the non affected partner) had to be balanced against respect for the independence of the partner with HD. The paper by Scerri (2015) who interviewed eight carers of HD partners describes the worries linked to the dependency needs of their partners as contributing towards what is called the ‘direct constant presence’ of HD. This anxiety might be linked to a variety of concerns where in small and major ways the HD affected person seeks to behave independently and self reliantly free from the anxious concern of their partner. Incidents like managing independently in the home, driving a car,

going on cycle trips and long walks could all be the focus of this struggle being enacted. Moreover, this particular couple dynamic could be reversed with the carer seeking to find independence from their partner and evoking concern in the HD affected partner. The carer's need for recuperative or 'me-time' was sometimes the reason for this reversal.

As the course of the illness unfolded, participants in the study revealed how the tension between togetherness and separateness was renegotiated as symptoms became more difficult to manage. A significant point of change occurred around the time that residential care needed to be considered for two of the couples. For these two couples, anxious concern for the HD partner was given second place to anxious concern for children resulting in the carers in these two families ejecting their partners from home using different legal means and, thereby, establishing an emotionally attenuated and a physically distant relationship. For the third couple where residential care was not wanted, it is likely that the physical separation heightened anxious concern in the non affected partner.

The effects of HD on couple intimacy: the roles of carer and being cared for.

Intimacy whether emotional or physical is, for many, a key and valued component of their adult partnership. Five of the seven couples in this study gave clear evidence that this aspect of their relationship had been severely disrupted by the impact of HD, a finding that was supported by results from the Literature Review. Of the two other couples, one intimated that a continuing sexual relationship was highly valued and the second offered no information in this connection. In particular, sexual problems are confirmed by a number of studies (O'Connor et al. 2008; Federoff et al. 1994; Reininghaus and Lackner, 2015; Kolenc et al. 2015; Baikie 2002) .

This study adds to this general finding by discovering the mixed feelings that can be associated with a loss of previously well functioning sexual relationships. While at least one partner from all five of the couples conveyed the distress and sadness that surrounded the relinquishment of an active sex life, it was clear that reactions to this loss could be more nuanced. One HD affected man talked of how the loss of sexual relationship made him feel second class. One would guess that he might have been speaking for two other men in the sample who, from the accounts of their partners, persisted in wanting intercourse long after

their partners were finding it enjoyable. By contrast four of the women in the sample, who were not affected by HD, talked about the relief that came from having ended sexual relations. Before this point was reached, their accounts point to the different ways in which intimacy could be linked with feeling emotionally hurt and/or physically harassed/pressurised and obligated into having intercourse. For all five of these couples it seemed as if one partner was to feel dissatisfied and the other satisfied whether they were having a sexual relationship or not.

A further tentative finding from this study was that the adopted roles of carer and being cared for ran in parallel with the ending of a sexual relationship. It was noticeable that the one couple who continued to enjoy a sexual relationship did not use or refer to the roles of carer and cared for in their interview while another couple explicitly linked the change in roles to the relinquishment of a sexual relationship. This linkage may suggest that for some couples there is recognition that caring and being cared for involves a desexualisation of their future relationship.

The move to residential care

Research concerning the move into residential care did not surface in the Literature Review but the accounts given by Helen, Sarah and Henry raise general points that may have wider relevance for a professional audience.

When comparing the accounts given by all three it is clear that the circumstances surrounding admission to residential care evoked contrasting emotional reactions. At the time of admission Henry was resisting the advice of professionals to have Tess admitted to a nursing home while Sarah and Helen were desperate for their partners to leave home. An understanding of these different responses is helped by having some awareness of the nature of the couples' relationships before HD entered their lives and, in Sarah and Helen's cases the immediate family context.

Henry described a very tight almost fused bond with Tess which was maintained throughout her illness. Some of this may be explained by both their backgrounds which were traumatic. Tess's life before she married Henry had been dominated by illtreatment and abuse of various kinds. Henry's parents' marriage had been characterized by domestic violence which elicited

in him a protective response towards his mother. From this it is possible to speculate that an element of their attraction to one another was a rescuer/victim dynamic which was reinforced by Tess developing HD. From Henry's account it is clear that he gained great fulfillment in looking after Tess to the point that contributions from professionals in the medical and social care system were either ignored or were seen as being uncaring and negligent.

Sarah and Helen's dilemmas in finally having to give priority to the interests of their daughters over those of their husbands were exceptionally difficult to resolve. The strength of attachments to both partners and children lay at the heart of the growing crises in both families. It is also telling that as Sarah tried to balance concern for her husband alongside protecting her daughters' interests she felt that no one agency would get involved and help her manage the conflict. Perhaps, this is an example of institutional defences at work (Menzies Lyth 1960) where referral on to another agency is used as a defence against tangling with an anxiety inducing problem?

When reading the accounts of Helen and Henry it is striking how both talk in the present tense about events in the past. One of the characteristics of individuals who are classified as 'unresolved' within the attachment theory classification system is that they show momentary lapses into talking in the present tense during interview and it is suggested that this is an indication of unresolved issues from the past either in relation to abuse or significant loss (Lyons-Ruth & Jacobvitz 1999). For example:-

Henry (C7):-

"The thing is, I had a full time job looking after her...and the bad thing about this was then when we were put on the radar, I never went and seen a doctor and they virtually said, who are you? You shouldn't be doing this on your own. Well I am. This is what I do. I don't ask for help.

Helen (C6) also resorted to the present tense:-

"He said, I can't stay there, if I stay there, I am going to commit suicide. And I said, Mark, I said, that is completely up to you. I have tried as hard as I can and I can't do it anymore. You have stayed at home with me and the girls for as long as you physically could and we cannot

have you at home any more, and so this is your life now. And this is where you live. I will come to see you...”

In fact Henry used the present tense far more than did Helen but, given that both had been bereaved in the recent past, it is possible that their use of the present tense confirms that both were preoccupied with mourning their partners.

5.2 An appraisal of whether the aims of the study have been achieved.

This study was exploratory in several respects. Its main aim was to explore how a diagnosis of HD impacts on couple relationships and, in pursuing this aim, the open ended questioning style adopted in individual and joint interviews was employed to understand ‘the lived experience’ of participants. In so far as the open ended questioning approach attempted to stay close to the experience of participants, the methodology allowed for themes to emerge from the data using TA. However, in addition to an inductive approach to the data, the analysis adopted a deductive approach through the application of attachment theory. A further aim of the study was to explore whether it might be possible to develop a modified version of FANIM so that it might be applicable to couple interaction.

Taking each of these elements of the study in turn, the following tentative conclusions are made. With regard to emergent data some of the findings parallel those made by other studies but have, in some areas, added to them.

1. The first overarching theme-Knowledge of the family history of HD- broadly overlaps with findings from other studies.
2. The second overarching theme- Talking or not talking about HD- between partners and within the nuclear and extended family-offers ways of conceptualising how that knowledge is handled which had not emerged from earlier literature searches. The analysis tracks how knowledge is handled within families first by the couples themselves and then on an intergenerational axis (involving parents and children) and on a cross generational axis involving members of the extended family.

The different ways that family members communicated or refused to communicate about HD touch on important questions about how individuals live with knowledge about progressive

mental and physical decline while at the same time needing to get on with everyday life. The analysis in this study suggests that the psychological defences of denial and avoidance may be necessary in order to free family members to lead fulfilled lives. Conscious awareness of HD and the importance of talking about it may only become salient (Etchegaray 2011) at particular times.

This is a challenging thought in a culture which equates positive mental health with the capacity to describe lived experience in coherent verbal ways. Frank (2013) has challenged this view suggesting that the expectation that people should always be encouraged to find words to match their experience is misplaced. Freud's notions of primary and secondary experience, Bion's theory of thinking which seeks to explain the move from preverbal experience to verbal/symbolic thinking via the mechanism of containment, and attachment theory's equation of adult security with narrative coherence, are all examples of theories within the counseling/psychotherapy fields that can be read as equating positive mental health with the ability to give coherent accounts of personal experience. Other mental states involving denial, avoidance or incoherent narratives are, on this reckoning, felt to be less healthy. In opposition to this view, Frank claims that some people may be faced by situations that are so traumatic that they may be 'beyond words' or may produce 'chaos' narratives' which are jumbled and confused stories reflecting the jumbled and confused mental states of the narrators. Such responses in his view are understandable in the circumstances and not pathological.

It is possible that my own emotional responses in the course of the research work illuminated Frank's identification of chaos. A number of events coincided at a point early in the fieldwork which lent weight to my growing awareness of the pain and distress that confronting the reality of HD can generate and made me appreciate the role denial and avoidance may take in helping families function on a day to day basis.

When the second couple rejected the opportunity to meet with me in a joint interview, having permitted me to interview them individually, I noted, at the time, that it felt like a 'real slap in the face'. I was quite thrown by this unexplained rejection, in part, because I thought I had established a good enough relationship with both partners. In attempting to understand this decision I concluded that discussing the couple relationship was for them a 'no-go area'; they might be able to talk to me separately about HD but focusing on their experiences together in

a joint interview was asking too much. The frustration was that I could not be sure about this judgment; I did write inviting an explanation for the withdrawal but I received no reply. In the background to this experience I also had the experience of interviews with the first couple which had impressed on me the stresses and losses associated with HD's arrival into family life and the courage it took to talk about them.

At about the same time I attended a meeting in a residential setting where an array of HD patients, their families and carers were present. Around a large lounge area sat or lay an array of people which, to my eyes, appeared to represent the complete HD illness trajectory. Sitting to my left were a young family where the mother found it difficult to control her chorea form movements as she sat in a chair. At her feet sat her two young, at risk, daughters colouring in books, seemingly oblivious to the adult discussions going on around them. By the door was a lady lying in a special bed whose body was fixed in a rigid, dystonic, position characteristic of the later stages of the illness (Quarrell 2008). Other seats were occupied by adults, some of whom were in intermediate stages of the illness. My hope when attending this meeting was that I might recruit further couples to my study but I was disappointed. I was told by two carers that they would be willing to participate in the study but that it would be too disturbing for their HD partners.

I can recall driving home from this meetings feeling stirred up by the experience. Coupled with the rejection by the second couple to engage in a joint research interview, I felt that, at best, I was being naïve and, at worst, arrogant and cruel to think that a research focus on the emotional life of HD couples was an ethically justified activity. I was being told explicitly by some couples that research interviews could be experienced as unwanted and emotionally destabilizing.

While I was wrestling with these experiences I had research supervision at the university. As I described them to my two supervisors and, in particular, recalled the dilemmas faced by children and their parents as they come to terms with genetic risk, I began to cry quite uncontrollably. I was hugely embarrassed at this happening and have since thought long and hard about the experience.

Frank's idea of a chaos narrative that cannot be put into words has helped me understand my response in that supervision session as being, in part, a counter-transference response to my

fieldwork-I couldn't find words to express what I was feeling and felt chaotically out of control. Within contemporary psychoanalytic theory and practice, counter-transference refers, not to the way that Freud viewed it-an enactment of the analyst's pathology triggered by something in the patient's material-but an unconscious communication from the patient through the processes of projection and identification. (An analogy would be the way an attuned parent picks up the distress of a child who cannot find words to communicate their discomfort).

Frank's way of understanding this phenomenon came in part from his reflections on his own experience of coping with testicular cancer. He claimed he was able to identify with the chaotic experience of his research subjects because he had been in analogous situations himself. I, too, have had cancer-in my case of the prostate-and, at the point of my being distressed in the supervision session, I was within weeks of coming to the end of two years of treatment and I was anxious about follow up blood tests.

So while it is conceivable that my distress had nothing whatsoever to do with my research work, it is possible that my vulnerability at the time made me particularly sensitive to the vulnerabilities of the HD families I was getting to know and that my distress, to some degree, echoed the chaos and confusion that HD families keep at bay through the defences of denial and avoidance. If this is so, then I came to appreciate more fully than before what I was asking research participants to talk about and share in future interviews and to respect the decisions of those who refused to participate.

The possibility that transient states of mind marked by chaos and confusion are not signs of pathology but are understandable and perhaps inevitable reactions when facing a degenerative illness raises several questions about mental functioning. It implies that individuals, couple and families struggling with serious illness operate on different levels of mental functioning.

On the one hand, there is a requirement to get on with the ordinary business of living when it is no longer possible to hold onto pre-illness assumptions about bodily integrity and the predictability of one's experiential world. Toombs (1987), writing about the phenomenology of illness, claims that there are five features that characterize the experience of chronic illness. These are the loss of wholeness-the body can no longer be taken for granted; the loss

of certainty-the effects of illness can be unpredictable in their timing and influence: the loss of control-the patient loses control of how the body operates; loss of freedom to act-life choices become restricted and, finally, loss of the familiar world-the patient becomes isolated from established social activities. Confronting the magnitude of these losses will be overwhelming for many, so that it is not surprising that maintaining a positive approach to life will require the mobilization of a series of defences, particularly of avoidance and denial such that some good qualities of living can be maintained. On the other hand, the limitations of illness, as described by Toombs, will be encountered in different ways. They may be consciously anticipated so that losses are prepared for but it is also likely that apprehension of what illness portends will catch those affected in unexpected ways. Psychological defences will be breached and anxieties that were put to one side will become salient (Etchegaray 2011). The study reveals some of the ways that confronting or avoiding knowledge about HD was managed by seven couples and how the implications of this knowledge may have permeated the research encounter.

3. The third overarching theme- Negotiating the transition from a pre-illness to an illness dominated relationship.

There were few studies in the Literature Review that looked at how couples managed their relationships over time with most focusing on couple and family functioning at particular points in time. This meant that accounts of dynamic processes within families and between couples were in a minority. This third theme makes a contribution to redressing this gap. In formulating this theme, concepts drawn from attachment theory are implicit. The concept of distance regulation at both physical and emotional levels underpins the three sub themes.

In relation to the first sub theme, the physical and emotional distance is linked to balancing anxious concern with a need to respect independence and, while it is perhaps not surprising that this balance is typically worked out between the non affected partner having to struggle with allowing their HD partner to remain independent for as long as possible, it was also true that the dynamic could work in the reverse direction particularly when the non affected partner needed time away from the carer role in order to recuperate; the bid for 'me-time' as one participant put it.

The second sub theme elaborated on the complex feelings that could be felt in relation to the loss of sexual intimacy and the increased distancing that followed. It was striking that partners would sometimes hold different views about what it felt like to end an intimate sexual relationship. Where men were HD positive and symptomatic, the evidence was that they generally resisted the ending of sexual intimacy whereas their partners felt relief. In this context it is important to recall that one of the neurological complications of HD can be sexual disinhibition. There was also some evidence that the ending of a sexual relationship was linked with taking on the roles of carer and cared for, which raises interesting questions about whether these two roles are essentially desexualized, akin to the sexual taboos that are often linked to nurse roles.

The third sub theme exploring the moves into residential care. The physical distancing that this move entailed was traumatic in all three families and was resisted in very different ways. In two families it was the male HD partners who resisted the evictions from home whereas in the third it was the male carer who resisted relinquishing his ill wife to the care of a nursing home.

One of the aims of the study was to consider the idea that a diagnosis of HD can be understood as an attachment threat to a couple relationship. The evidence from the interviews in this study is that this is too simplistic a way of understanding the impact of an HD diagnosis. While it was the case that many participants described hearing the diagnosis as ‘devastating’, most families had been making adjustments to the disease for months and, in some cases, years before the formal diagnosis was given. Moreover alongside the feelings of shock and dismay that a positive diagnosis evoked, there could be some relief that an explanation for previously unexplained symptoms was now available which strengthened rather than weakened the attachment bond to the affected person.

However, the crises that surrounded the admissions into residential care did in all three families represent an attachment threat for the evicted spouse. For the two women who had to use legal processes to evict their partners they decided that their need to protect the welfare of their children took precedence over the need to continue to care for their husbands. These two painful situations were stark reminders, among many others, that a focus on couple interactions could not be separated out from wider family concerns.

4. The study looked at the possibility of modifying the FANIM method to be used with couples. FANIM innovatively introduces the idea of the defended subject and I explored whether this could be applied to thinking about couples as a shared system. While some aspects of FANIM could be usefully deployed in this research-for example, the importance of looking at defences in research participants and researchers-there were limitations on exploring the application of FANIM rigorously and there were several reasons for this. In the first place there were not enough couples to complete the full set of interviews (separate individual interviews followed by a joint interview). Only two of the couples completed this programme. In addition there was a tension in my interview style between, on the one hand, wanting to gather information that would have contributed to formulating a psychosocial/defended couple as required by FANIM, and, on the other hand, finding that, as my confidence grew, my interviewing technique developed into using more open ended and fewer questions/prompts, thus allowing participants to set more of their idiosyncratic stamps on interview content. This more open approach meant that the questions and prompts listed in Appendix 2 were rarely used in the order they appear there. However, they became internalized as a range of subject areas that I attempted to cover in all interviews. With the size of sample involving ten individuals, six of whom comprised three couples Thematic Analysis emerged as the most appropriate method to identify inductive and deductive themes. There were a number of aspects to my way of conducting the analysis which are interesting in hindsight. Among the strongest impressions were:-

- It was not my experience that I gave equal attention to all items in the data set as is advocated in the TA literature. Recruitment to research projects about sensitive topics can be challenging and uneven. There was a five year interval separating the first interview from the last and the last four couples were interviewed in a three month window. As a result, Couples C1 and C2, plus the couple who withdrew from the study, received considerable attention and thought at the outset of the fieldwork which informed the later reshaping of the study. In this way, in depth analysis commenced from the beginning of commencing the interviews and this thinking influenced later contacts. For example, I came to realise that participants wanted to speak more urgently about their current difficulties in managing HD than the order of the prompts in the interview schedules allowed for. Moreover, as has been mentioned, a professional transcriber was employed to help with the last four interviews. The transcription process involves an interpretation of what is said on tape and, while each

transcription was checked for accuracy, it is possible that because an additional transcriber was involved there may be an unevenness in interpretation.

- I sensed that the initial stage of becoming immersed /familiar with the data can be described as a creative form of playing. I noted that “close reading of the interview transcripts brings the memories of the actual interviews flooding back. There is a curious feeling of approaching and revisiting something very familiar alongside a sense of looking at the experience with fresh eyes. The result is that some preconceptions are confirmed, others are modified and new insights are suggested. As the process of coding takes shape on the page I am struck by the randomness of what catches my attention enough to note down. Coherence only slowly emerges from what I pick out which can vary from specific words or phrases, details on interviewees’ lives, impressions that I form at different stages of the interview, my own feelings, strategies of the interviewees to engage with particular topics e.t.c” (Note written for Mental Health Research Group Seminar 10.9.2016).
- The second and third stages involved separating out the initial codings and seeing whether they clustered in particular ways. I undertook this process by writing the provisional clusters in pencil in the margins of the hard copy. Using pencil and rubber enabled me to amend and develop the clusters that I saw emerging. It seemed to me that this process involved a move from relative randomness to increased coherence. It seemed also to involve moving away from idiography to thinking about the sample as a whole and drawing on theory to do so (For example, the importance of emotional and physical distance regulation). As this process advanced (supported by supervision) it was increasingly possible to envisage how emerging themes related to established research.

5.3 Limitations of the study

The limitations of this study will be considered under the headings of sample, research methodology and research data analysis.

5.3.1 Limitations of the sample. It was intended to recruit a purposive sample but, in fact, the sample became a convenience sample reflecting the difficulties of recruiting participants. The sample remained purposive to the extent of only involving HD families but the initial aim of interviewing both partners was modified to allow couple experience to be accessed through the involvement of one partner. Recruitment of couples to the study proved difficult reflecting, in part, the problems of couples openly sharing the challenges of living with HD and the challenge of sharing sensitive information about intimate matters with an unknown researcher. Only two couples completed the full set of interviews and with four of the couples data was provided by one partner only so that recall of events in family life was made from different and partial points on the illness trajectory. Whether these differing vantage points on couple functioning enable general claims to be made about the whole group is then open to question as is extrapolating general claims from this study to the wider population of HD couples.

5.3.2. Limitations of the research methodology. 1) The research methodology adopted in this study evolved during the fieldwork stage to take account of participants' agendas and priorities. In particular, adherence to the order in which questions were listed in the interview schedules was relaxed in favour of allowing participants to determine the order in which topics were addressed. Had this approach been adopted at the first two interviews it is possible that the responses may have been more spontaneous and closer to participants' lived experience.

2) A strong finding from the study was the discovery that asking couples to share in face to face interviews their thoughts and feelings about living with HD may have been too challenging for some while others appreciated the opportunity the research offered to talk together with a third party. While I was given ethical clearance to conduct the study and conducted it with care, this finding raises the possibility that other means might be employed to obtain the same evidence. Methods of enquiry that preserve anonymity and/or do not expose partners to each other's thinking might prove more acceptable for some couples and/or offer complementary data.

5.3.3 Limitations of research data analysis. The data produced in this study has been generated and analysed by one person with the help of supervision which raises the objection that the findings are not adequately validated by some independent means. The 'hermeneutic circle' can be criticized for producing findings that conform the observer's presuppositions and theories and lacks the validation provided by a second researcher or by the feedback from participants.

Chapter 6. Conclusion

Unlike research studies that are commissioned from the outset to answer a particular problem, this study grew out of a number of converging interests. The author's interest in the psychology of ageing led to a collaboration with colleagues in the National Health Service who had established a specialist service to support families contending with early onset dementia (EOD). The possibility emerged to develop a research study in that service exploring how early onset dementias were experienced and managed by couples which anecdotal and research evidence suggested bore high levels of stress and the the major costs. A Scoping Review was undertaken at a time of major financial cutbacks in the NHS resulting in the dedicated service for EOD families being axed. One outcome of the Scoping Review was a helpful working relationship with staff at the Huntington's Disease Association who agreed to support the study which, in the absence of an NHS base, was refocused to explore how couples managed HD.

An important premise behind this study was a belief that the dearth of research into how couples experienced HD represented a void which affected couples would welcome being filled by a research study. There is a well established recognition that many qualitative research studies have given voice to marginalized members of the community including those with serious degenerative diseases.

Findings from this study suggest that this early premise needs qualification. In the form that this study took-inviting couples to be interviewed in separate and joint interviews-the evidence was that recruitment was difficult. While recruitment to studies investigating sensitive personal and family issues is recognized as being difficult, several instances of affected individuals stating that being interviewed jointly was too difficult , suggests that, when facing a serious illness, there are limits to the extent to which couples can jointly share, in a very brief time limited research context, their thoughts and feelings about living with HD. This conclusion is supported by the greater ease with which individuals came forward to talk about their experiences.

The findings from the study also add weight to the significance of sharing or not sharing information about illness in a variety of different couple and family situations. The study identified that dilemmas about communicating and sharing knowledge about the disease

ranged across two dimensions. There was sometimes an intergenerational challenge about who should be told what about the fact or the risk of inheriting the illness. Some families were faced with the paradox of needing to protect a grandparent from being told about the illness while at the same time actively sharing news of the illness with the younger generation so that decisions can be taken about family planning. It may be that these families were in a unique position in medical and social history as they attempted to move from a culture where eugenic prejudice was still detectable to one where advances in medical knowledge allow for a more rationally based approach to family planning.

The difficulties in sharing knowledge about HD were also identified within the extended family when HD was confirmed. Some families who were coming to terms with the implications of HD for themselves realized that there was an ethical case for talking about the implications of HD for the members of the extended family who might be at risk of carrying the faulty gene. Their experience was that this news was difficult to convey and was often denied leading to inter-family rifts.

In developing these themes, the material builds on and is complementary to the work of other researchers. As such it contains within it the evidence of couple and family processes which can be moulded into other formats-lectures, training materials, journal papers e.t.c-which may be of value to the training and continuing professional development of professionals working in this field. It is also possible that it contains evidence that will be of benefit to service users contending with HD.

More generally, it is fair to say that this study, being based on an exploratory premise and limited to a small sample, raises further questions. For example, it invites a consideration about the forms of information and support which best help couples at the point of diagnosis and how can HD families can be better supported and helped to plan for residential care.

I believe that the accounts in this report are true to the experiences I shared with the three couples and four individuals who were generous in volunteering to be interviewed. I believe that all did so because, at the times they were interviewed, they had a need to share their stories with me, partly because some were explicit about wanting their stories to be heard so that others in similar circumstances might benefit from them but, also, because, without stating it, they realized that talking about their experiences was of benefit to them. These twin

motivations create an ethical and a psychological problem which I hope have been addressed sensitively in this thesis and in any future publications or training materials that follow on from it. The ethical challenge is that there is a need to report in a respectful way on the most private and sensitive aspects of participants' couple and family lives in ways which do not compromise their interests. The psychological problem, which is embedded in the participants' stories, is whether I and others can bear to listen to and talk about personal stories that involve serious existential dilemmas. These involve losing control of one's mind and body, of facing inevitable death and living with the consequences of passing on the illness to children. This thesis charts the ways the participants, and I as researcher, have jointly attempted to face and consider how these challenges continue to be resolved.

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Appendices

Appendix 1: Scoping Review Documents

1. Ethics approval

Southampton

Eo4/Aug 2010/ v1.1

Chris Vincent
60, North Shore Road
Hayling Island
PO11 0HN

10 September 2010

Dear Chris

Ethics Submission No: SoHS-ETHICS-2010-022
Title: Scoping review for main study

I am pleased to confirm **full approval** for your study has now been given. The approval has been granted by the Faculty of Health Sciences Ethics Committee.

You are required to complete a University Insurance and Research Governance Application Form (IRGA) in order to receive insurance clearance before you begin data collection. The blank form can be found at <http://www.soton.ac.uk/corporateservices/rgo/regprojs/whatdocs.html>

You need to submit the following documentation in a plastic wallet to Dr Martina Prude in the Research Governance Office (RCO), University of Southampton, Highfield Campus, Bldg. 37, Southampton SO17 1BJ:

- Completed IRGA Research Governance form
- Copy of your research protocol/School Ethics Form (final and approved version)
- Copy of participant information sheet
- Copy of SoHS Risk Assessment form, **signed**
- Copy of your information sheet and consent form
- Copy of this SoHS Ethical approval letter

Continued overleaf

Building 67
Faculty of Health Sciences, University of Southampton, Highfield Campus, Southampton SO17 1BJ United Kingdom
Tel: +44 (0)23 8059 7979 Fax: +44 (0)23 8059 7900 www.southampton.ac.uk/healthsciences

Your project will be registered at the RCO, and then automatically transferred to the Finance Department for insurance cover. **You can not begin recruiting until you have received a letter stating that you have received insurance clearance.**

Please note that you have ethics approval only for the project described in your submission. If you want to change any aspect of your project (e.g. recruitment or data collection) you must request permission from the Ethics Committee and RGO (students should discuss changes with their supervisor before submitting the request to the Ethics Committee).

Yours sincerely


Professor Sue Lister
Chair, FoHS Ethics Committee
t: +44 (0)23 80 597959
e: sml@eoton.ac.uk
f: +44 (0)23 80 597900

2

Building 67
Faculty of Health Sciences, University of Southampton, Highfield Campus, Southampton SO17 1BJ United Kingdom
Tel: +44 (0)23 8059 7979 Fax: +44 (0)23 8059 7900 www.southampton.ac.uk/healthsciences

2. Invitation letter.

Date

A SCOPING REVIEW TO HELP DEVELOP A MAIN STUDY WHICH WILL EXAMINE THE IMPACT OF A DIAGNOSIS OF YOUNG ONSET DEMENTIA ON PATIENTS AND PARTNERS

AN INVITATION TO TAKE PART

Dear.....

Further to my talk onabout the research I am undertaking I would like to invite you both to take part in the scoping review to help me develop a research study which will explore the impact of a diagnosis of young onset dementia on couple relationships.

Your cooperation is entirely voluntary and will involve you and your partner agreeing to take part in a joint interview at a time and place to suit you. I anticipate that these consultations take between 60 to 90 minutes and, in order to help me hold on to your advice, I am proposing that the consultations will be audio-taped.

One aim of the National Dementia strategy is to provide an effective and early diagnostic service to all patients and their families. This study is being undertaken as a contribution towards that end. It is a key assumption behind this project that an improving and effective service will be offered if health care professionals have a good understanding of the experiences of patients and their partners.

I attach a Participant Information Sheet, dated August 25th 2010, which explains in fuller detail the background to this project and questions related to your possible involvement in the Scoping Review.

If you would like me to answer further questions you can contact me on:-

02392 358010

Yours sincerely

Christopher Vincent

M.Phil student
Faculty of Health Sciences

Draft: August 25th 2010
Ethics No:2010-022.

3. Participant Information Sheet

Participant Information Sheet STUDY TITLE

A SCOPING REVIEW TO HELP DESIGN MAIN STUDY: TO EXPLORE THE IMPACT ON PATIENTS AND THEIR PARTNERS OF A DIAGNOSIS OF YOUNG ONSET DEMENTIA

Researcher: Christopher Vincent

Ethics number: 2010-022

I would like to invite you and your partner to take part in my research study. This information sheet contains information about the study which will help you decide whether you wish to participate. If anything is not clear or there other matters that you would like clarifying, please do not hesitate to raise them with me.

Please read this information carefully before deciding to take part in this research. If you are happy to participate you will be asked to sign a consent form.

What is a scoping review? It is a consultative exercise in which service users (which includes patients and members of their families) are asked their advice and opinions in the development and design of a research project.

What is the research about?

The overall aim of the main study will be to explore how hearing about a diagnosis of early onset dementia affects the relationship of patient and partner. Knowledge about the impact of the diagnosis on this important relationship in the lives of patients may help health care professionals develop and improve existing services to families.

What are you being asked to help with?

The main study will be based within a specialist NHS service providing diagnostic and on-going care to patients with an early onset dementia and there are three broad lines of questioning that I am proposing to explore in this future study. I would value your advice whether these questions are appropriate.

The three broad areas are:-

First, I am proposing to ask couples about the adaptations and changes they found themselves having to make following the diagnosis, comparing the quality of their relationship in the year before and after diagnosis.

Second, I am proposing to explore how adaptations couples have to make in the present connect to underlying strengths in their relationship. I am assuming that an

understanding of what brought couples together and how they got on early in their partnership will reveal these underlying strengths.

Third, I am proposing to ask couples what they rate as the positive and negative aspects of the NHS diagnostic service. I will also ask them about any gaps in service provision.

In addition to these questions, I would also value your advice about the most effective ways of communicating the results from the main research study to health care professionals, service users and the general public. I will also like to ask your views about whether you think there is a role for patients to be involved in analysing research data.

I hope that on balance talking about your experiences will be helpful and that you may derive satisfaction from knowing that, in the long run, the purpose of researching this subject is to contribute towards an improved NHS diagnostic service. However at any stage you have every right to withdraw or to terminate the interview without explanation. The manager of theCentre has agreed to offer you both follow-up discussions to the one with me if you feel this would be needed or would be helpful.

My background and position.

I am a student in the Faculty of Health Sciences at the University of Southampton undertaking a post graduate research degree. My background includes developing training programmes for health care staff.

Why have you been approached to help?

You have been approached to take part through the Alzheimers Society/ Huntington's Disease Association where senior staff have agreed to cooperate with this project and have given me permission to contact you.

What would cooperating mean for you both in practical terms?

If you agree to help with this scoping review, I would ask to interview you both together at the offices of or at your home. There will one interview lasting between 60 to 90 minutes which, to help me hold on to the information given by you, will be audio-taped with your consent. In the event that you elect to hold the interview away from your home I am pleased to pay your travel expenses at a rate of 40p a mile. I am sorry that I cannot recompense other costs incurred.

I am conscious of asking a lot from you in asking for help with my study. Because of this and wishing to hear your opinions uninfluenced by involvement with other studies in the same area, I am excluding from this scoping review any couples who

might be involved currently or recently with providing their views on service provision.

What if one or both of you changes your mind?

At any point before or during the interview you can ask to terminate the discussion without giving any explanation and that decision will be respected. If you wish we can consider whether you would like the interview to be reconvened at a later time. If you decide to end the interview, I will then ask whether you would like any taped material to be destroyed or would still allow the information you have already shared to be used in the study. Information will only be retained and used if you both agree. Any decision to withdraw from the scoping review will not affect your treatment or care from theCentre.

Is the information given treated in confidence?

The material you share with me will be treated with the utmost respect and will be suitably anonymised in the ways it will be subsequently used. The method for doing this will be as follows:-

1. The audio-tapes will be kept in a locked filing cabinet in my office.
2. Their content will be transcribed onto computer file with all identifying personal references changed or removed. Thus names and employment will be changed and addresses removed.
3. The means by which this disguised material will be linked back to you will be through a unique reference number which will link your details with the tape and transcription. A record linking you to your reference number will be a master file kept securely and separately from the tapes and transcribed copies in a separate locked cabinet.

The standard I will adopt to secure confidentiality is that no one apart from yourselves and me would be able to identify you from the transcribed and edited texts. The requirements of the Data Protection Act will cover the handling of this material. All data will be stored for ten years according to the regulations of the University of Southampton.

The transcribed material may be used within the final thesis I will be writing as part of my research degree. It may also be used in subsequent publications-professional journals, conference papers, book chapters-or in training with your consent.

Has this scoping review the backing of The University of Southampton?

This scoping review has been approved by the Ethics Committee of the Faculty of Health Sciences at the University of Southampton.

Is this study in receipt of funding from a sponsoring body?

This project is, as yet, not in receipt of any funding from a sponsoring body. Should this change during the course of the scoping review I will let you know.

How do we proceed if we have a complaint to make?

If you have any concern or complaint about this scoping review I hope you will be able to discuss this with me in the first instance. However, if you would like to direct your complaint directly to the University you should contact Susan Rogers, Head of Research and Enterprise Services, Faculty of Health Sciences, University of Southampton, Building 67, Highfield, Southampton, SO1 7 1BJ, tel: 02380597942, email: s.j.s.rogers@soton.ac.uk. If you remain unhappy and wish to complain formally Susan Rogers can provide you with details of the University of Southampton Complaints Procedure.

I hope this information sheet provides you with all you need to know about the scoping review and that you will be able to make an informed decision about helping. Attached to this Sheet are two Consent Forms, one for you to retain, that you will need to complete if you wish to take part in the scoping review. Many thanks for your help so far.

Christopher Vincent

M.Phil student/Faculty of Health Sciences

4. Consent Form

CONSENT FORM

Title of Project: Scoping Review to help design main study: to explore the impact on patients and their partners of a diagnosis of young onset dementia.

Name of Researcher: Christopher Vincent

Please initial box if you agree

1. I confirm that I have read and understand the Participant Information Sheet dated August 25th 2010 for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. ☐
2. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason and without my legal rights being affected. ☐
3. I agree to our consultations being audio-taped ☐
4. I understand that my words may be used in an academic thesis and professional publication, subject to the levels of disguise and protection of identification explained in the Participant Information Sheet dated August 25th 2010. ☐
5. In the last year I have not been involved in a research study asking me to comment on health or other professional services. ☐
6. I agree to take part in the above study. ☐

Name of Participant:
Name of Participant:

Date:
Date:

Signature:
Signature:

Name of Researcher:

Date:

Signature:

When completed: I for participant & I for research file
Consent Form draft August 25th 2010: Ethics No.2010-022

5. Interview Schedule

INTERVIEW SCHEDULE

Scoping Review to help design main study: to explore the impact on patients and their partners of a diagnosis of young onset dementia.

1. I thank Mr and Mrs (Ms)..... for agreeing to take part in this consultation and remind them of our having met at.....where I had explained the purpose of the main study and how this scoping review fits into it. I respond to any questions they have about matters that are not clear or have been forgotten. I confirm that they have read the P.I.S and have signed the Consent Form.
 2. I check out that they recall our meeting and, on the basis of what we now understand, remain happy about continuing with the interview. I also remind them that either of them can terminate the interview at any stage if they are not feeling comfortable when we can discuss what actions are then needed.
 3. If they remain content to proceed I remind them of their agreement to the interview being audio-taped and ask their permission for the machine to be switched on
 4. I then ask them whether they can say a little bit about themselves-how long they have been together, whether they have children themselves, what sort of jobs/careers they have or had, how their link with theCentre began.
 5. I then remind them that my main purpose in meeting with them is to gain their views about the appropriateness of asking certain specific questions of other couples in order to understand the impact of the diagnosis and diagnostic experience on these couples' partnerships.
- **What problems/opportunities might it raise for couples if I ask them about the adaptations that they had to make in their relationship during and subsequent to the diagnosis?**
 - **Would it help if I asked them to describe their relationship in the 12 months before the diagnosis was made and ask them to compare it to their relationship in the subsequent 12 months?**
 - **Do you think it is appropriate to ask couples about how events around the time of the diagnosis affected their relationship?**
 - **In trying to understand how couples have responded to a current event in their lives, do you think it would be helpful to understand how they got on together in the first twelve months of their relationship?**
 - **Do you think that couple relationships are at their strongest then i.e. in the first twelve months?**
 - **Do you foresee any problems in asking these questions?**

- **Do you think there are any other questions that it might be important to ask to more fully understand the impact of a diagnosis of early onset dementia on a couple's relationship?**
 - **Do you think that exploring the impact on couples might help improve services?**
 - **Do you think it is useful to ask participants in the study how they experienced the service they received from professionals in the health service, social services and voluntary agencies?**
6. I then remind my consultees that I would like to ask their views about how best the results of this project can be communicated.
 7. If no particular views are forthcoming I will ask whether for them particular communicative media have been helpful in understanding the consequences of the illness-books, films, TV, internet articles, newspaper articles, talks, conferences, others?
 8. I then ask whether they think there would be any value in service users, including themselves, being involved in analysing the data emerging from this project.
 9. Finally I will ask consultees to reflect on the interview and to let me know how they are feeling about it as we come towards an ending. Depending on their replies, I will suggest that they might like to share their experience with.....(voluntary agency staff member)
 10. Before leaving I will thankfor their cooperation.

Draft 5/8/10
Ethics No.2010-022

Appendix 2: Main Study

1. Ethics approval

Your Ethics Submission (Ethics ID:5701) has been reviewed and ap... <https://www.outlook.soton.ac.uk/owa/?ae=Item&a=Open&t=IPM.N...>

[Reply](#) [Reply All](#) [Forward](#)

Your Ethics Submission (Ethics ID:5701) has been reviewed and approved

ERGO [ergo@soton.ac.uk]

To: [Vincent C.M.](#)

Submission Number: 5701
Submission Name: Exploring the impact of a diagnosis of Huntington's Disease on couple relationships
This is email is to let you know your submission was approved by the Ethics Committee.

You can begin your research unless you are still awaiting specific Health and Safety approval (e.g. for a Genetic or Biological Materials Risk Assessment)

Comments

1.Dear Christopher. Thank you for your thorough and thoughtful response to the queries from the ethic committee reviewers. We are happy with your amendments and explanations. Wishing you good luck with this interesting piece of research.

2.I too am happy with the amendments

[Click here to view your submission](#)

ERGO : Ethics and Research Governance Online
<http://www.ergo.soton.ac.uk>

DO NOT REPLY TO THIS EMAIL

1 of 1 15/01/2014 16:05

2. Advertisement in the HDA Newsletter

EXPLORING THE IMPACT OF A DIAGNOSIS OF HD ON COUPLE RELATIONSHIPS

I am a research student in the Faculty of Health Sciences at Southampton University where I am about to start a qualitative study looking at how a diagnosis of HD impacts on couple relationships. Knowledge about the impact of the diagnosis on this important relationship may help health care professionals develop and improve existing services to families. It is in line with the National Dementia Strategy which aims to improve services to patients at the point of diagnosis and the focus on couple relationships recognizes the fact that this is a key relationship in maintaining the stability of family life..

I have already undertaken pilot work and am at the stage of having been given ethical clearance to undertake the main study.

How can members of the HDA help?

I am now seeking the help of four volunteer couples where one of the partners was diagnosed with HD no longer than ten years ago and no more recently than two years ago. I will ask each couple to share their experiences in two separate individual interviews and one joint interview suitable spaced apart. I found that in my earlier pilot work these interviews were best undertaken in volunteers' own homes and I propose that we adopt the same arrangements for this next stage.

The interviews will be audio-taped and their content will be subject to the University's strict requirements to ensure that confidential information is properly respected and that the requirements of the Data Protection Act are fully observed.

If you are interested to share your experiences I will send you full details about my study which will explain in more detail its rationale, the sort of questions I will be asking and the safeguards built into the procedures to safeguard your interests.

How to contact me

I can be contacted by email at:-

XXXXXXXXXXXXXXXXXXXXXXX

XXXXXXXXXXXXXXXXXXXXXXX

Thank you for reading this note and for considering my request

Christopher Vincent

3. Invitation Letter

Date

A STUDY TO EXPLORE THE IMPACT OF A DIAGNOSIS OF HUNTINGTON'S DISEASE ON COUPLES

AN INVITATION TO TAKE PART

Dear.....

Further to my talk/our discussion onabout the research I am undertaking I would like to invite you and(both) to take part in my study.

Your cooperation is entirely voluntary and will involve you and your partner agreeing to take part in three interviews in your own home at a mutually convenient time. As you will see from the Participant Information Sheet the interviews will comprise two separate and one joint interview and I anticipate that each one will take between 60 to 90 minutes. The interviews will be audio-taped.

One aim of the National Dementia strategy is to provide an effective and early diagnostic service to all patients and their families. This study is being undertaken as a contribution towards that end. It is hoped that an improved and effective service will be offered if health care professionals have a good understanding of the experiences of patients and their partners.

I attach a Participant Information Sheet, dated September 27th 2013, which explains in fuller detail the background to this project and questions related to your possible involvement in the study.

If you would like me to answer further questions you can contact me on:-

02392 358010

Yours sincerely

Christopher Vincent

M.Phil student
Faculty of Health Sciences

Draft: September 27th 2013
Ethics No.5701

4. Participant Information Sheet

Participant Information Sheet

Study Title: Exploring the impact of a diagnosis of Huntington's Disease (HD) on couple relationships

Researcher: Chris Vincent

Ethics number: 5701

Please read this information carefully before deciding to take part in this research. If you are happy to participate you will be asked to sign a consent form.

What is the research about?

This study is an in-depth exploration of the impact on and responses of couples where one partner has had a medical diagnosis of Huntington's Disease. This illness has far reaching and stressful consequences which place a burden on the adult couple and, if there are children, their capacity to parent. In order to strengthen and support this relationship it is important that professionals in front line services are attuned to the stresses experienced at the point of diagnosis. This study aims to improve our understanding of the experience of couples and, in doing so, will contribute to one of the aims of the National Dementia Strategy which is to improve diagnostic services to families.

My name is Chris Vincent and this study is part of my working towards an academic qualification. I am a mature student and have spent most of my career working as a clinician with couples who face challenging life situations. I developed an interest in the particular challenges posed by Huntington's Disease having interviewed one couple contending with this problem at an earlier phase of this research project.

My approach will be to understand as far as possible you and your partner's experience told in your own way and, thus, my questions will be general and designed to open up a conversation. Within this broad remit, I have some general areas that I would like to cover. For example, I will be interested to know about how your shared and separate experiences of facing and managing stressful life events in the past have influenced your more recent experience of managing HD. I will also be interested to understand how HD has changed your relationship and influenced the quality of family life.

This research is not externally funded and is sponsored by the University of Southampton.

Why have I been chosen?

I am grateful that you are considering participating in this study which involves interviews with four couples where the formal medical diagnosis took place no longer than ten years and no sooner than two years ago. This time gap ensures that some adjustments will have been made to living with HD but not so long ago that memories of the time around diagnosis will be lost.

What will happen to me if I take part?

Should you agree to take part in this study, you and your partner will be interviewed by me on three occasions. The first two interviews will involve me interviewing you separately. The purpose of meeting with you separately is to gain a full picture of you as individuals; for

example, a separate interview will help me gain a picture of your family background when growing up and to gain a picture of your life before forging a partnership together. The third interview will be a joint interview which will focus on your relationship together both before and after the Huntington's diagnosis was made so that I can gain a good understanding on how it has impacted on your lives and the kind of adjustments you have had to make. All three interviews will be audio-taped and will last approximately an hour and no longer than an hour and a half. In the final part of the joint interview I will turn off the audio-tape machine and invite you to share with me your feelings and thoughts about the sequence of interviews as a whole.

Are there any benefits in my taking part?

There may be no benefit to you in taking part. However, the aim of the research is to generate knowledge and understanding which will be fed back in appropriately anonymised form to front line professionals in the hope that diagnostic services will be improved. If you wish I will keep you informed of the ways that this feedback happens.

Are there any risks involved?

I will be asking you to share personal experiences with me (whom you have not met) and this may feel challenging and risky in prospect. My experience is that pursuing this sort of enquiry in a way that respects the sensitivity of the matters being discussed can be experienced as beneficial by those who are generous enough to take part.

Will my participation be confidential?

The material you share with me will be treated with the utmost respect and will be anonymised as follows:-

1. The audio-tapes will be kept in a locked filing cabinet at the University of Southampton. Only I will listen to them.
2. Their content will be transcribed onto computer file with all identifying personal references changed or removed. Thus names and employment will be changed and addresses removed.
3. The means by which this disguised material will be linked back to you will be through a unique reference number which will link your details with the tape and transcription. A record linking you to your reference number will be a master file kept securely and separately from the tapes and transcribed copies in a separate locked cabinet.

The standard I will adopt to secure confidentiality is that no one apart from yourselves and me would be able to identify you from the transcribed and edited texts. The requirements of the Data Protection Act will cover the handling of this material. All data will be stored for ten years according to the regulations of the University of Southampton.

The transcribed material may be used within the final thesis I will be writing as part of my research degree. It may also be used in subsequent publications-professional journals, conference papers, book chapters-or in training with your consent.

Has this study the backing of The University of Southampton?

This research has been approved by the Ethics Committee of the Faculty of Health Sciences at the University of Southampton.

The research is supervised by Dr Joanne Brown and Professor Peter Coleman.

What happens if I change my mind?

At any stage of your participation in the research you have the right to withdraw without in any way compromising your interests. You do not need to explain the reasons for wishing to withdraw and, as I am not involved with your medical care or involved with any other organisation, your care will not be adversely affected.

How do we proceed if we have a complaint to make?

If you have any concern or complaint about this scoping review I hope you will be able to discuss this with me in the first instance. However you may decide to approach the University directly. You should contact Martina Prude, Head of the Governance Office, at the Research Governance Office (Address: University of Southampton, Building 37, Highfield, Southampton, SO17 1BJ ; Tel: +44 (0)23 8059 5058; Email: rgoinfo@soton.ac.uk . If you remain unhappy and wish to complain formally Martina can provide you with details of the University of Southampton Complaints Procedure.”

I hope this information sheet provides you with all you need to know about the study and that you will be able to make an informed decision about helping.

Many thanks for your help so far.

Chris Vincent

5. Consent Form

CONSENT FORM (September 27th 2013)

Study title: Exploring the impact of a diagnosis of Huntington's Disease on Couple Relationships

Researcher name: Chris Vincent

Study reference:

Ethics reference: 5701

Please initial the box(es) if you agree with the statement(s):

I have read and understood the Participant Information Sheet (September 27th 2013) and I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily.

☐

I agree to take part in this research project and agree for my data to be used for the purpose of this study.

☐

I agree to our interviews being audio-taped.

☐

I understand my participation is voluntary and I may withdraw at any time without my legal rights being affected.

☐

I agree that my words may be used in an academic thesis subject to the levels of disguise and protection of identification explained in the Participants Information Sheet (September 27th 2013)

☐

Data Protection

I understand that information collected about me during my participation in this study will be stored on a password protected computer and that this information will only be used for the purpose of this study. All files containing any personal data will be made anonymous.

Name of participant (print name).....

Signature of participant.....

Date:.....

Name of researcher:.....

Signature of researcher:.....

Date:.....

6. Individual Interview Schedule

INDIVIDUAL INTERVIEW SCHEDULE

1. Can you tell me something about yourself? (prompts) Background, family, perhaps the big events in your life that you feel have influenced you?
2. What was it like growing up in your family? (prompts) happy/unhappy childhood, place in sibling order?
3. How did you view your parents' relationship? (prompts) warm or conflicted, has their relationship had a bearing on what you expect from your partnership?
4. Were there any challenges to your sense of security when growing up and how did you experience them? (prompts) separations, illnesses, hospitalisations
5. Can you say something about the quality of your teenage and early adult years? (prompts) smooth transition to adulthood or troubled/difficult, education/leaving home, employment aspirations
6. Before you met and married/partnered X, what was your experience of previous relationships?

September 27th 2013 Version
Ethics no. 5701

7. Joint Interview Schedule

JOINT INTERVIEW SCHEDULE

1. Can you say how you met and what it was about your relationship that attracted you to one another? (prompts) what positive qualities did you see in him/her and what positive qualities do you think he/she saw in you?
2. Before HD became a factor in your lives, could you communicate your neediness to your partner and was that need met? Can you give an example?
3. What happened the other way around i.e. when your partner expressed emotional neediness and wanted support from you? Can you give an example?
4. Can you tell me how and when it was that you realise HD was something you/your partner would suffer from? (prompts) family history, early symptoms, genetic test.
5. Can you tell me what that time was like and how it affected you:-
 - (1) as an individual? (prompts) mood, outlook on life, genetic test, were you able to talk about HD together, support or otherwise of family/friends/neighbours, role of HDA, involvement of health (GP and specialist) and social services.
 - (2) as a couple can you tell me what changed in your relationship? (prompts) changes for the better/for the worse, how compares to examples given earlier re asking for and giving support, impact on finances/housing.
6. Can you tell me when you received a formal diagnosis of HD and what that was like? (prompts) relief/shock, quality of services received/emotional responses to professionals
7. Can you tell me how coming to terms with HD affected you in your role as parents or as parents to be? (prompts) genetic tests, birth control, impact on child(ren)'s behaviour.
8. Can you tell me how coming to terms with HD has influenced your attitude(s) to the present and future?

September 27th 2013
Ethics no. 5701

Appendix 3: Extension of sample to include one case from the Scoping Review and four individual participants

1. Ethics Approval

From: ERGO [ergo@soton.ac.uk]
Sent: Wednesday, March 25, 2015 9:58 AM
To: Brown J.C.
Subject: Ethics ID:14451 has been reviewed and approved

Submission Number :14451
Submission Name :Exploring the impact of a diagnosis of Huntington's Disease on couple relationships (Amendment 2)
This is email is to let you know one of your student submissions has been reviewed and approved by the ethics committee.

They can begin their research unless they are still awaiting specific Health and Safety approval (e.g. for a Genetic or Biological Materials Risk Assessment)

ERGO : Ethics and Research Governance Online
<http://www.ergo.soton.ac.uk>

DO NOT REPLY TO THIS EMAIL

Appendix 4: Ethical approval to use participant's diary entries as part of the data set

1. Ethics Approval

From: ERGO [ergo@soton.ac.uk]
Sent: Friday, December 11, 2015 1:08 PM
To: Brown J.C.
Subject: Ethics ID:18540 has been reviewed and approved

Submission Number :18540
Submission Name :Exploring the impact of a diagnosis of Huntington's Disease on couple relationships (Amendment 3)
This is email is to let you know one of your student submissions has been reviewed and approved by the ethics committee.

They can begin their research unless they are still awaiting specific Health and Safety approval (e.g. for a Genetic or Biological Materials Risk Assessment)

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