University of Derby

The development and validation of a scale to measure the impact of Huntington's Disease on the quality of life of Spousal carers.

Aimee Aubeeluck

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Preface:

Motivation for Research.

Huntington's Disease (HD) is a rare condition that has been under-researched by the medical professions and psychologists alike. There is a clear lack of psychological literature on the subject of HD and furthermore, there are no adequate QoL scales available for use by spousal carers. The development of a HD specific QoL scale (HDQoL-C) for this special population, brings together theoretical constructs and practical application in order to produce a user-friendly QoL measurement for spousal carers of HD patients.

Aims and Objectives.

Huntington's Disease (HD) is a dementia that is genetically inherited as an autosomal dominant trait with complete life-time penetrance (Zakzanis, 1998). The majority of the HD literature focuses on the patient and those 'at-risk' of inheriting the HD gene within the family, rather than the spousal carer, who has been referred to as the 'forgotten person' in the HD family (Kessler, 1993). Research into the experience of the HD spousal carer has established that carers experience a number of unique obstacles within their caregiving role (e.g. Hans & Koeppen, 1980). However, such research is both sparse and limited and there is a clear need to establish methodically the factors that impact upon the HD spousal carer's situation and ultimately their quality of life. This research is aimed at systematically investigating the factors that enhance and compromise the lives of HD spousal carers by utilising the theoretical construct of quality of life (QoL). My exploratory studies provide

evidence that spousal carers of HD patients have specific difficulties in maintaining their QoL whilst continuing in a primary care-giving role. As such, they prepared the way for the development of a disease-specific QoL measure for HD carers (HDQoL-C). The objective of this scale is to quantify the caregiving experience in HD in order to implement and assess therapeutic interventions.

"From week to week and month to month he experienced more and more difficulty controlling what he called "this damned nervousness." And when he became nervous, he had trouble with his temper. For years he had been a gentle and understanding husband. Now he seemed to get irritated with his wife and children at the slightest pretext. Jean scolded him for being too rough on the kids. For the first time in their married life, he shouted at her. In several occasions he hit her. She wondered if Ian had problems at work that he did not wish to discuss. She spent dark and gloomy hours worrying about what had come over him. Had she let him down somehow? Was there another woman? Had he taken to drinking secretly? There was no sign of any of it, but her husband was clearly a changed man"

(excerpt taken from Phillips (1982). Living with *Huntington's Disease, a book for patients and families, p4).*

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Abstract.

The purpose of this thesis was to systematically investigate the factors that enhance and compromise the lives of Huntington's Disease (HD) spousal carers by utilising the theoretical construct of quality of life (QoL). Three exploratory studies provided evidence that spousal carers of HD patients have specific difficulties in maintaining their QoL whilst continuing in a primary care-giving role. Study 1 provided preliminary evidence that spousal carers of HD patients and health care professionals would value a disease specific QoL scale that could be used to evaluate spousal carers' objective and subjective QoL. Study 2 established that spousal carers of HD patients often experience loneliness, a need to escape and a unique sense of loss whilst trying to adequately care for their loved ones and maintain some form of QoL for themselves. Study 3 provided further evidence that QoL is compromised in many ways and is an issue for HD carers. The carers in this study often neglected their own needs as the caregiving role and disease processes 'took over' their lives as well as the life of their HD affected spouse. The findings of these three studies prepared the way for the development of a disease-specific QoL measure for spousal carers of HD patients. Validation of the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) was carried out in study 4. The newly developed scale was found to have good internal consistency, test-retest and face validity. It is hoped that the HDQoL-C will be used in the future to quantify the caregiving experience in HD in order to implement and assess therapeutic interventions.

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I would like to thank my wonderful family for their support and encouragement throughout the duration of my studies and also Heather Buchanan, a fantastic supervisor and fabulous friend.

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To Megan and Chloe: It's all for you...

List of original publications, conference posters and invited presentations resulting from this research.

Journal Articles.

- Aubeeluck, A., & Buchanan, H. (In preparation) Validation of a quality of life measure for spousal carers of Huntington's Disease patients.
- Aubeeluck, A., & Buchanan, H. (In preparation) The Quality of Life experienced by spousal carers of Huntington's Disease Patients: Qualitative findings from focus groups.
- Aubeeluck, A., & Buchanan, H. (In Press) Capturing the Huntington's Disease spousal carer experience: A Preliminary investigation using the 'Photovoice' method. (see appendix I)
- Aubeeluck, A. (In press). The Huntington's Disease Quality of Life Battery for Carers (HDQoL-C). (see appendix II)
- Aubeeluck, A (2003). Spousal caregiving in Huntington's Disease: An Issue for health psychology. *Health Psychology Update* 12(4)19-25. (see appendix III)

Conference presentations.

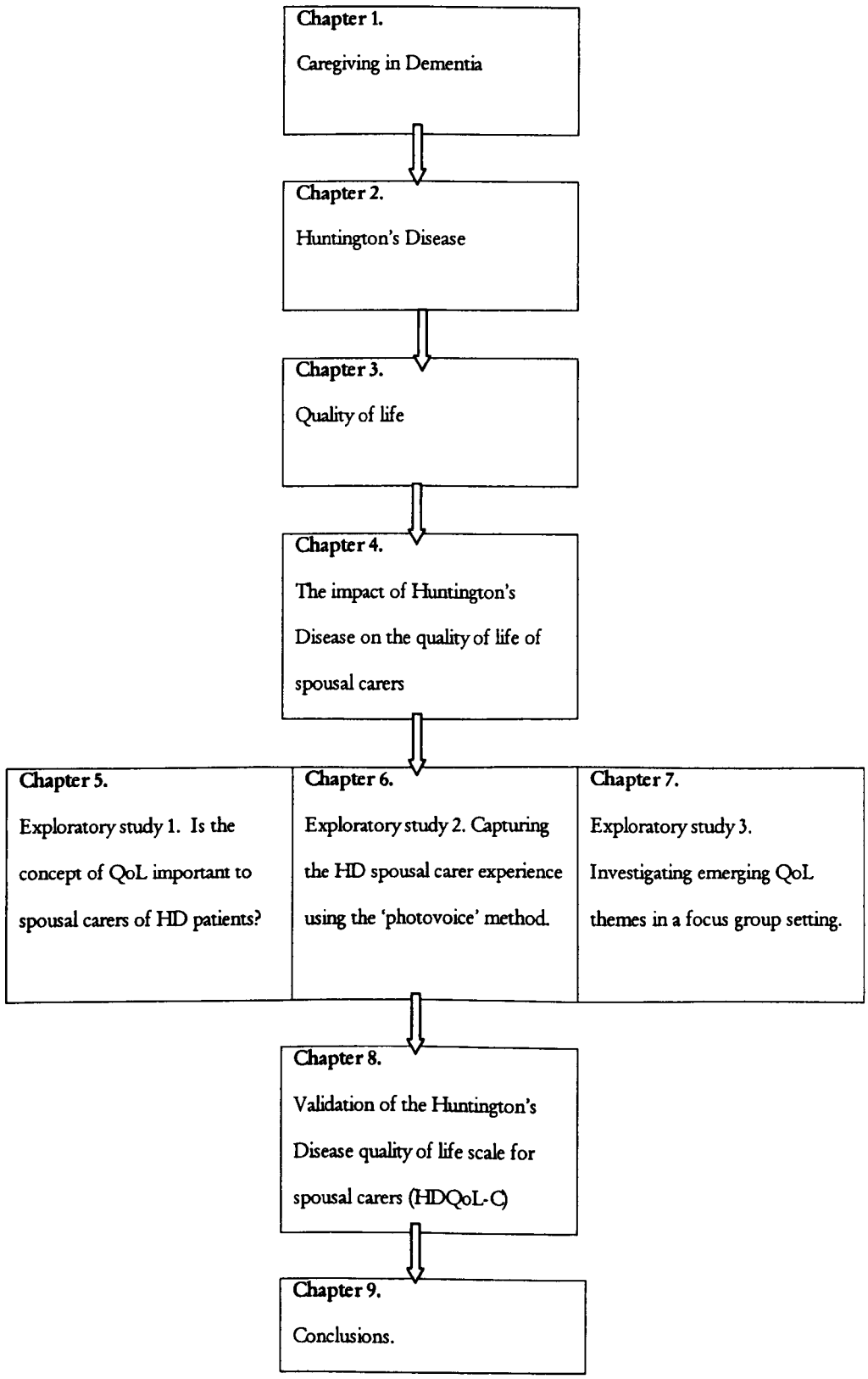
- Coulson, N.S, Aubeeluck, A., Buchanan, H., & Semper, H., Rooney, E. (2004). 'Exploring the provision of social support within a computer-mediated Huntington's Disease support network' Poster presented at 18th Annual Conference of the European Health Psychology Society, Helsinki.
- Coulson, N.S, Aubeeluck, A., Buchanan, H., & Semper, H., (2004). Participation in an online Huntington's Disease support network: who participates and what do they talk about? Paper Presented at BPS Health Psychology Annual Conference, Edinburgh.

- Aubeeluck, A. & Buchanan, H. (2003). Developing a quality of life measure for spousal carers of Huntington's Disease patients. Poster Presented at Health Psychology Annual Conference
- Aubeeluck, A., & Buchanan, H. (2002). Huntington's Disease Family Carers: Compromising quality of life in order to care? Poster Presented at Health Psychology Annual Conference

Invited presentations.

- "Exploring interventions for family carers of Huntington's Disease patients". Workshop run for Devon Branch of the Huntington's Disease Association (May 2004).
- "Huntington's Disease & its impact on the quality of life of patients and their families". Guest Lecture presented in the Department of Psychology, University of Nottingham (April, 2004).
- "Spousal caregiving in Huntington's Disease: implications for quality of life". Talk presented at the Huntington's Disease Association AGM (December 2003).
- "Quality of Life in Huntington's Disease Carers". Talk presented to the Devon Branch of the Huntington's Disease Association (May 2002).
- "Exploring the effects of caring: Huntington's Disease and Quality of Life of family carers". Talk presented at the Huntington's Disease Association AGM (September 2001)

Overview of Chapters.



1. Spousal Caregiving in Dementia.

This initial chapter introduces the concept of spousal caregiving in dementia placing particular focus and discussion on the theoretical background of family caregiving and the impact that the caregiving role can have on the life of a spouse.

2. Huntington's Disease.

This chapter outlines the clinical characteristics of Huntington's Disease (HD). The history, neurology, neuropsychology and neuropsychiatry of HD will be described. Furthermore, the epidemiology and genetics of HD will be highlighted and issues surrounding genetic counselling and presymptomatic testing will be critically discussed.

3. Quality of life (QoL).

For the purpose of this research, QoL is for the most part utilized as a psychological tool that can be used to evaluate outcome measures rather than a theoretical concept. However, in this chapter, the theory behind the construct of QoL will be discussed. QoL will be defined, giving a brief history of the construct and an attempt will be made to conceptualise the construct in relation to this current research. QoL will then be discussed as a concept and its use as a psychological tool within Health Psychology will be addressed.

4. The impact of Huntington's Disease on the quality of life of spousal carers.

In this final introductory chapter, family, and specifically spousal caregiving, in HD is discussed. The motivation for the research and research questions will also be outlined.

5. Exploratory study 1: The importance of QoL to spousal carers of HD.

In this initial study the domains and facets of the Comprehensive Quality of Life Questionnaire for Adults (ComQol-A5) (Cummins, 1997), a well validated generic QoL measure, were rated by using a likert type scale in order to achieve two main objectives: (1) to examine the relevance of the ComQol-A5 domains and facets to the perception of HD Spousal Carers and (2) to consider the development of any additional facets & remove less relevant facets. Carers and health care professionals were also asked to write down any issues that they felt were pertinent to their QoL as primary carers of HD patients. Analysis of the ratings data revealed that participants perceived every facet as either very important or extremely important to their quality of life. Further analysis of the qualitative data established 18 sub-themes relating to quality of life which clustered into four final themes of: Professional Issues in HD, Personal Wellbeing in HD, Practical Issues in HD and Emotional Wellbeing in HD.

6. Exploratory study 2: Capturing the HD spousal carers' experience using the 'Photovoice' method.

The objective of the second exploratory study was to investigate the factors salient to the QoL of family carers in more detail. In order to gain insight into the complex role of the carer, visual representations of QoL and corresponding written information were gathered using 'Photovoice'. 'Photovoice' was employed as an opportunity for individual participants to explore the concept of QoL by photographing and giving written reflections on specific QoL issues surrounding their care giving and HD. Using content analysis, nine manifest themes were identified

and tentative latent inferences were made in relation to these themes. Moreover, although some positive issues did emerge, these were minimal compared to the negative impact that HD had on carers' overall QoL.

7. Exploratory study 3: Investigating emerging QoL themes within a focus group setting.

The findings from studies 1 and 2 provided evidence that QoL is both a concept that is relevant to HD spousal carers and further that their QoL is greatly impacted upon due to their caregiving role. The purpose of a third exploratory study was therefore to further investigate previous findings within a larger sample of spousal caregivers and to provide a clear framework for designing a HD specific QoL measure for spousal carers. Therefore, in this third study the conceptual framework of the initial draft of the Huntington's Disease quality of life Battery for Carers (HDQoL-C) was tested through the use of 6 semi-directed focus groups. Analysis of the focus group data supported the identification of the four themes identified in study 1 (Professional Issues in HD; Personal Wellbeing in HD; Practical Issues in HD; Emotional Wellbeing in HD) and further identified the new theme of HD specific issues. All observed themes and sub-themes were integrated into the existing ComQoL-A5 (Cummins, 1997) to generate a HD specific QoL measure for spousal carers (HDQoL-C).

8. Validation of the Huntington's Disease quality of life scale for spousal carers (HDQoL-C).

The objective of this pilot study was to validate the HDQoL-C with spousal carers of HD patients. As such, 87 full time spousal carers of HD patients completed the pilot

questionnaire. Factor Analysis established the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) as a multidimensional and psychometrically sound disease-specific and subjective QoL assessment tool which incorporates the individual's physical health, psychological state, level of independence, social relationships, personal beliefs and relationship to salient features of the environment. The HDQoL-C demonstrates good internal consistency, test re-test reliability and congruent validity for use with spousal carers of HD patients.

9. General Conclusions and Future Directions.

This final chapter discusses the development of the Huntington's Disease Quality of Life Battery in relation to the extant dementia and HD caregiving literature. Future directions for the use of the HDQoL-C are also discussed as is continuing research into family caregiving in HD within the field of Health Psychology.

Chapter 1.

Family Caregiving in Dementia- a brief overview

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Introduction:

The dramatic ageing of the UK population (Office of National Statistics, 2004) has focused attention on to chronic illnesses such as dementia and their societal impact. Chronic illness does not only impact upon the patient themselves, but also requires that families and sometimes friends take on the role of informal carer (Carers National Association, 1997). The tasks and burdens associated with caring are often numerous and varied, frequently changing across the course of an illness (Brown & Stetz, 1999). Tasks can range from running errands and provision of emotional support to more practical aspects such as assisting bathing, feeding or other activities of daily living and the management of disruptive behaviours. Families and friends may have to juggle their lives in order to make room for these care responsibilities and their related stressors whilst continuing in other substantive familial and social roles such as being a spouse, parent, friend or employee. Caregiving responsibilities and the need to balance such roles can often lead to caregiver distress and many family carers become the "forgotten person" (Kessler, 1993) who is left to struggle with their own physical and mental health concerns by themselves (Bookwala et al, 2000; Coon et al, 1999).

1. Caring for a family member with a dementing illness.

Models of caregiving.

The World Health Organisation define dementia as "... the global impairment of higher cortical functions including memory, the capacity to solve the problems of day to day living, the performance of learned perceptuo-motor skill, the correct use of social skills and control of emotional reactions, in the presence of gross 'clouding of consciousness'. The condition is often irreversible and progressive." (WHO, 1986, p 2). Therefore, dementia is a syndrome or grouping of syndromes which can manifest itself in various combinations and in which the individual has very little awareness of what is happening to them. Dementia usually starts with relatively slight impairments but can progress to a point where all the skills of communication and self-care are lost. As such, the symptoms and nature of the disease change over the course of time with stages being labeled as mild, moderate or severe and disease progression being dependent on both internal and external factors (Jones & Meisen, 1996). In end stage dementia, patients will require full-time care which can be highly demanding and prolonged (Wallsten, 1993). Furthermore, care is generally provided by partners, who are often elderly and unprepared for the physical and emotional demands placed on them (Braithwaite, 1996).

Most current caregiving research is built on stress and coping or stress-process models in order to understand the impact of caregiving (e.g. Lazarus and Folkman, 1984; Haley et al, 1987; Pearlin et al, 1990; Nolan et al, 1996). Lazarus and Folkman (1984) argue that an individual may engage in a number of coping strategies in order to reduce the adverse

emotional state associated with the appraisal of (perceived) stress. These fall into the two categories of problem-focused and emotion-focused coping. The goal of both strategies is for the individual to control their level of stress. In problem-focused coping, people try to short-circuit negative emotions by taking some action to modify, avoid, or minimize the threatening situation, i.e. the individual attempts to change the situation in order to reduce its threat. In emotion-focused coping, people try to directly moderate or eliminate unpleasant emotions without actually trying to change the situation. Examples of emotion-focused coping include rethinking the situation in a positive way, relaxation, denial, and wishful thinking. A number of more specific categories are subsumed within these two broad categories as outlined below:

Illustration 1.1. Examples of Problem-Focused and Emotion-Focused coping from Lazarus and Folkman's (1984) Stress-Coping Model.

Problem-Focused Coping	Emotion Focused Coping
Confrontive Coping	Distancing
Seeking Social support	Self-control
Planful problem solving	Positive reappraisal
Accepting responsibility	Escape/avoidance

Both types of coping can occur simultaneously (Folkman & Lazarus, 1980) and the success of any effort depends on the individual involved and the nature of the challenge. In general, problem-focused coping is the most effective coping strategy when people have realistic opportunities to change aspects of their situation and reduce stress. Emotion-focused coping is most useful as a short-term strategy. It can help reduce one's arousal level before engaging

in problem-solving and taking action, and it can help people deal with stressful situations in which there are few problem-focused coping options. The model focuses on coping processes that can be used to manage or reduce aversive states. However, it is also based on the assumption that an event is not stressful unless the individual perceives it to be stressful (Lazarus, 1966; Tomaka et al, 1997).

This model, when applied to caregiving, places emphasis on the carer to re-interpret their situation as non-threatening. For carers who are already overburdened, this added pressure may be overwhelming. Furthermore, within a chronic caregiving situation, there may be very little realistic opportunity to change any aspect of their situation. Indeed, for dementia carers it is probable that the stressors they encounter are likely to increase throughout the duration of their caregiving role as the burden 'mounts up' over the course of time (George & Gwyther, 1984, cited in Duijnstee, 1994). Moreover, Kneebone & Martin (2003) note that research based on Lazarus & Folkman's (1984) model is severely limited in its capacity to inform the clinician as much research fails to incorporate measures of strategies which may aid the clinician in identifying what coping works, for what problem and when. Therefore without further identification of specific stressors against which to measure such coping strategies, the Lazarus and Folkman (1984) model may not prove very useful in terms of implementing successful interventions.

Haley, Brown & Levine's (1987) stress process model places emphasis on stress as a relationship between the caregiver and the environment and includes stressors, appraisal, coping responses, social support and adaptational outcomes. Haley et al propose that each

component of the model is assessed independently for a clear conceptual understanding of the relationship between stressors and caregivers. The model is outlined below:

Illustration 1.2. Haley, Brown & Levine's (1987) Stress Process Model (adapted from Coon et al, 2003).

Stressors are described as environmental and psychological influences that are problematic for the carer. Therefore, how the carer appraises a stressful situation, copes with a stressor and uses available resources combines to form the conceptual description of mediators. Variability in caregiver outcomes (i.e. the degree to which the caregiver successfully adjusts

to their caregiving role), is believed to be influenced by the differing mediators that are used by each individual caregiver. This model is useful as it conceptualizes the physical and psychological well-being of the carer as 'outcomes'. Therefore, it can be used to measure the efficacy of skills training interventions and provides evidence of the problem solving skills that carers utilize in order to achieve specified outcomes. However, evaluations of its predictive utility should be taken with caution due to the small sample size and variety of carers used in its validation.

Pearlin et al (1990) developed a framework that allows the demands and resources of the caregiver to be clearly identified. They distinguish four domains: background and contextual factors; stressors; mediators of stress; and outcomes. The stressors are themselves divided into three types: primary stressors, directly connected with providing care; secondary role strains such as those caused by the conflicting demands of caring, work and family; and secondary intra-psychic strains, including the impact of caring on self-esteem. They have further developed an interview schedule to pick up the major factors within each of these domains. This particular framework accounts for many influential factors such as culture and life history. Other mediating factors include resources for coping with social, economic and internal stresses. The schedule provides a clear system for outlining a carer's situation and can also be used to suggest relevant interventions. The model is outlined below:

Illustration 1.3. Pearlin's stress-process model of stress in carers of those with Alzheimer's disease (adapted from Pearlin et al, 1990).

Alternatively, Nolan et al (1996), have produced a descriptive, stage-based, longitudinal model of the caregiving process, based on findings from interviews with carers who were supporting people with varying degrees of Alzheimer's dementia. The six stages of this model are: building on the past; recognising the need; taking it on; working through it; reaching the end; and a new beginning. The basic stress-appraisal-coping process (recognition and appraisal of stress; appraisal of resources and ways of coping; taking action and evaluating action) takes place within each of the stages. This model can be helpful in prompting the clinician to consider the dynamic 'career' of the caregiver and to tailor any intervention to the appropriate stage of the process. However, the scale was developed using only Alzheimer's carers as participants and although this population of carers will of course experience some of the same stressful situations as carers of patients with other dementing illnesses, there are differences between dementias which may make a scale based purely on the Alzheimer carers experiences not as useful in other dementing illnesses such as Huntington's Disease (HD).

Bourgeois et al (1996) argue that a lack of theory-driven research has generated methods, design and measurement strategies that are disjointed from one another and lead to conflicting results. As such, the complexities of dementia caregiving suggest that new models or modifications to existing models are required in order that we better understand the dynamic nature of caregiving and the relationship between the carer, care recipient and biopsychosocial variables. Researchers, service providers and policy makers are finding it increasingly important to have useful frameworks to conceptualise effectively the caregiving needs of caregiving families. In the current climate, it is also important to understand how the caregiving process is influenced by non-traditional family units (e.g. divorced and blended families), families with divergent cultural views of the meaning of caregiving and the obligations that such cultural perspectives may bring. Different causes of stress among differing caregiving populations may require new models or conceptual frameworks with which to work in order to incorporate any unique aspects of the caregiving role.

In response to such issues, new models have been emerging within the caregiving literature that attempt to address the multifaceted nature of the caregiving role. For example, Montgomery and Kosloski (1999) have developed a marker framework which looks at the developmental phases of the caregiving process as determined by the needs of different kinds of caregivers. They argue that there is no single generic carer role but rather that caregiving emerges from prior role relationships and is integrated with other roles. Therefore, caregiving is a dynamic process that unfolds over time with variable durations for different caregivers. This process involves seven key 'markers' that mark significant shifts in the caregiving process (e.g. defining oneself as a caregiver, seeking assistance and formal

service use). Furthermore, distinct groups of carers such as spouses or adult children are assumed to experience markers differently and at different intervals leading to alternate caregiving trajectories that require specified and unique types of service or support. Caron et al (2000) also place emphasis on phases of caregiving rather than stages of disease and caregiving is defined by the tasks and challenges faced by families along a caregiving continuum which ranges from prediagnosis, diagnosis, role change and chronic caregiving to the transition to alternative care and end of life issues. Such models may prove useful in the assessment of caregiver burden and distress as they take into consideration the multifaceted nature of the caregiving role and place it within the context of the stage of illness and the time spent caregiving.

Tools to Measure Caregiver Stress.

In assessing the degree of caregiver stress, it is important to distinguish between objective and subjective burden. Objective burden is the amount of additional practical support that must be undertaken by the carer in order to effectively care for the dependent person. Subjective burden is the emotional and cognitive reaction of the carer to their situation. The same amount of caregiving, in terms of practical workload (i.e. objective burden), may be experienced very differently by carers (i.e. subjective burden), depending on their personal interpretation of their situation (e.g. Lazarus and Folkman, 1984). There are approximately 55 instruments that are available to measure caregiver stress/distress in some form and about half of these are specific to caregivers of Alzheimer's or Dementia patients (e.g., Caregiver Activity Survey, CAS; Davis, 1997; Caregivers Stress Scales; Pearlin et al, 1990).

The Zarit Burden Interview (ZBI) (Zarit et al, 1980) is probably the most well-known tool developed to measure caregiver distress. It is a 29-item self-report scale which measures subjective burden on a likert type scale from 'not at all' to 'extremely'. However, the sample size used to validate the scale was very small (n=29) and the psychometric properties and construct validity of the scale are not entirely clear (George & Gwyther, 1986, Zarit, 1990). Nevertheless, its face validity is good, and its acceptability to carers has meant it has become widely used.

Another example of such measures is The Caregiving Burden Scale (Gerritsen & Van der Ende, 1994), a well constructed and validated scale which draws the majority of its items from the Zarit Burden Interview. It uses two factors to measure subjective burden: relationship and personal consequences. It is a 13-item scale that requires answers on a 5-point Likert scale from 'disagree very much' to 'agree very much'. The scale was developed in The Netherlands with a first sample of 89 carers for people with dementia and a second sample of 42. It has sound psychometric properties and good construct validity. It is quick to administer and is acceptable to carers, although some may require help to make distinctions within the 5-point response scale. In contrast, the Caregiver Hassles Scale (Kinney & Stephens, 1989) and the Behavioural Assessment Scale of Later Life (BASOLL; Brooker, 1998) both assess the degree of objective burden and the subjective level of stress that different aspects cause the carer. Both scales are lengthy and detailed (which can be problematic for carers who are restricted time wise). However they do provide a thorough picture of the support that is being provided as well as the carer's reactions to each aspect.

The Caregiver Hassles Scale has good psychometric properties and the results distinguish stress in each of five domains such as hassle associated with assisting with the basic activities of daily living and hassle with the carer's support network. However, it does not tap stressors related to changes in the relationship *per se*. The BASOLL is primarily a behaviour scale and psychometric properties are not given for the component dealing with the reaction of the carer. However, it gives a rich individual picture and can provide useful clinical information on where to target an intervention to relieve stress.

The purpose of assessing a carer's situation is to gain an understanding that may help to prevent or to ameliorate stress-related problems, enabling the carer to continue providing care comfortably for their relative (assuming that this is what both would wish). On occasion, the intervention may also appropriately lead to the carer letting the relative have care away from the home. The basis of successful intervention therefore, lies in gaining an informed understanding of the individual case. As such, using models of caring may provide helpful ways of organising the information on care levels, stresses, satisfactions and ways of coping that is gained during assessment. Stress-process models, for example, demonstrate possible areas for intervention at the level of reducing primary or secondary stressors, working to address intra-psychic strain or improving mediators such as coping skills or social support. Zarit & Edwards (1999) suggest that following assessment, the clinician needs to consider both the appropriate treatment strategy and the appropriate treatment modality. The main treatment strategies include: the provision of information; assistance with problem-solving to manage stressors; and providing or identifying sources of emotional and practical support. The modalities include one-to-one counselling or therapy; family meetings; and

support groups. Although these may sound straightforward and almost common sense, in practice they may be complex because of the intertwined emotional and practical issues involved. For example, a person who attributes a relative's repetitive questions to a lack of attention, may require information about the nature of memory loss. However, they may be emotionally defended against the knowledge that their relative has dementia so getting the information across in a sensitive yet useful way can take time and be difficult to achieve.

A meta-analysis of research between 1980 and 1990 (Knight et al, 1993) suggests that psychosocial interventions and respite care delivered on an individual basis each produced a moderate beneficial effect on carer distress. Group psychosocial interventions showed a small positive effect and other health and social services had no consistent impact on carer distress. In a later review Cuijpers & Neis (1997) examined research between 1987 & 1993 and concluded that individual interventions have significant positive effects, support groups enable people to use services appropriately; but respite care has limited effects. Acton & Kang (2001) also conducted a meta-analysis of 24 published research reports and found that interventions had no effect of caregiver burden. They suggest that burden may be too global an outcome to be consistently affected by intervention and argue that better and more precise measures are needed to evaluate the effects of caregiver interventions.

Therefore, it is possible that the lack of successful intervention findings may be related to the lack of consistency in the models and assessment methods that researchers utilise. Although stressors and strains that are associated with dementia caregiving are well documented (e.g Davidson, 1997; Mittleman et al, 1995; Schulz et al, 1995) and theory driven stress-process models exist that can help to suggest suitable interventions, the

majority of interventions that are actually described in caregiver intervention research are not explicitly theory-based or theory-driven themselves (Coon et al, 2003). Furthermore, these models place emphasis on distress and burden with less attempt to measure more positive aspects of caregiving such as the development of personal growth or feelings of self-worth or general quality of life. Lim & Zebrack (2004) conducted a review of 19 caregiver studies from 1987 to 2004 and found predominantly negative terminology used within caregiver questionnaires and only one measurement that explicitly measured quality of life *per se* (and this is not disease-specific to carers of dementia patients (The Caregiver Quality of Life Index-Cancer scale; CQOLC, Weitzner et al, 1999).

However, despite such issues, stepped care models have been applied to the investigation of individual differences as well as to the tailoring of interventions to meet the unique needs of divergent groups of caregivers (see Davidson 2000 for discussion). When applied to family caregiving, the stepped care process has the flexibility to assume that not all carers need the same type of intensity of intervention and that interventions should be minimally intensive / intrusive for the carers themselves. Therefore, despite their shortcomings, these models still provide an essential guide to intervention continuation or alteration and as such are useful in assessing the efficacy of an intervention and minimizing the cost of health care.

The Impact of Caring for a family member with a dementing illness.

Family members play a leading role in homecare for the demented elderly. Shanas (1979), Cantor (1983) and Johnson (1983) argue that help from the 'informal network' (i.e. partners, children, other relatives, friends and neighbours) is the most important source of support. Caring for a family member with a progressive dementing illness appears to be unique in the challenges it creates for the family caregiver. Various studies demonstrate the personal, health and social impacts of dementia care (e.g. Clark & Bond, 2000) and the financial burden that it places upon the family (e.g. Harrow et al, 2002; Ory et al, 1999). Family carers of people with dementia more frequently experience burden and burnout (O'Connor et al, 1990; Almberg, Grafstrom & Winblad, 1997;), have poorer self reported physical and mental health problems (Gonzalez-Salvador et al, 1999; Keicolt-Glaser et al, 1989), somatic illness (Levin et al, 1984) and have compromised immune function (Keicolt Glaser et al, 1987, 1991; Vedhara et al, 1999, 2000). Furthermore, carers for individuals with dementia in comparison to carers of non-demented individuals report heightened levels of stress and morbidity (Eagles et al, 1987; Dura et al, 1990; Rabins et al, 1990) and also describe more employment complications, greater family conflict and more family leisure time constraints (Ory et al, 1999). These issues raise a number of concerns with regards to the importance of recognizing the needs of caregivers relative to non-caregivers. Furthermore, with regards to ensuring that service providers, researchers and policy makers identify the best way to assist carers by taking into account their differing needs in relation to stressors and the need for formal support to help maintain their physical and mental health (Coon et al, 1999).

Within a given family, one person usually takes on board the primary care role (Cantor, 1983). In the case where an individual needing care has a partner, the partner usually becomes the primary-caregiver (Jones & Meisen, 1996) with more women than men assuming the caregiving role (Horowitz, 1985; Orbell 1996). Children may also find themselves in the primary caregiving role, either because the patient no longer has a partner or because the partner has become unable to continue in their primary caregiving role. Primary caregivers who turn to their children for support tend to turn to daughters before sons, and female offspring are more likely to place themselves in a full time caregiving role than male offspring (Dwyer & Coward, 1991). Orbell (1996) argues that power differentials between men and women and social expectations that caregiving is 'naturally' a woman's role mean that the predominance of women in family caregiving cannot be explained on economic grounds alone. That is, societal expectations surrounding the role of women put them in a position where they feel a duty to care.

Extant research has also found that older caregivers, women, carers who are employed, carers who live with the care recipient and those related to the care receiver as a child or spouse experience greater distress in association with caregiving (George & Gwyther, 1986; Pruchno & Resch, 1989; Biegel et al, 1991; Bass et al, 1994; Schluz et al, 1993). These are troubling findings, as carers represent a larger labour force than the NHS and social services combined with some 1.5 million people providing care for more than 20 hours per week (Carers National Association, 1997). Furthermore, between one-fifth and one-third of carers have provided care for more than 10 years and a small number of carers are under the age of 18 years (Webb & Tossell, 1999).

Summary.

The need to support family caregivers of dementia patients is clearly evident with the extensive literature related to caregiver distress in dementia (e.g. Keady & Nolan, 1995; Williams et al, 1995). The insidious and changing nature of dementia means that the caregiver role is always evolving and creating new problems / challenges for carers, often over a long period of time. How carers cope and adjust to their ever changing role may depend on how stressors are perceived and interpreted (Lazarus & Folkman, 1984). However, it is important to note that for carers in a chronic caregiving situation, there may be very little realistic opportunity to change any aspect of their situation, therefore rendering stress-process theory somewhat limited in its application to real life situations.

It is also apparent that in order for effective interventions to be implemented, there is a clear need for a comprehensive theoretical framework which takes a holistic perspective of the carer within the context of their situation (Keady and Nolan, 1996). It has been suggested that a lack of consensus in the conceptualization and measurement of caregiving outcomes may be responsible for the lack of consistent findings within dementia caregiving outcome research (Cousins et al, 2002). Furthermore, as carer populations differ in the problems they encounter, appropriate assessment tools need to be developed to 'tap in' to the areas in which carers may be struggling. Although the research literature tends to emphasize the

negative aspects of caregiving, areas in which carers are coping well should also be identified so that support is put into place where it is most needed.

Chapter 2.

Huntington's Disease.

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Introduction.

Huntington's Disease (HD) is a chronic progressive dementia of the brain that causes movement abnormalities (i.e. chorea and dystonia), cognitive deterioration and affective disturbances (Folstein, 1989). Symptoms typically begin at around the age of 40, with wide variation. Patients become severely demented, motorially dilapidated, unable to care for themselves and eventually bedridden. There is no cure for HD, with the treatments available being purely palliative or purely experimental and death occurring on average 15-17 years after onset (Harper, 1996). HD is a genetic condition inherited as an autosomal dominant trait with complete lifetime penetrance. Therefore, each person whose parent has HD is born with a 50:50 chance of inheriting the gene.

HD is often characterised by progressive involuntary choreiform (dancelike) movements. However, many patients manifest behavioural changes before the onset of the movement disorder. Early symptoms include lack of concentration combined with short-term memory lapses, depression and changes of mood that may lead to aggressive and / or anti-social behaviour as well as slight choreic movements, stumbling and clumsiness (Harper, 1996). Later on in the illness, earlier symptoms become exacerbated and patients often experience other symptoms such as constant involuntary movements, difficulty in speech and swallowing, severe weight loss, emotional changes resulting in a fixed mindset, frustration, mood swings and depression (Quarrell, 1999). Sufferers who experience cognitive changes that result in a loss of drive, initiative and organisational skills (Martin, 1984) may therefore have difficulty concentrating on more than one thing at a time making them appear lazy and uninterested. Sometimes these types of behavioural problems, rather than the physical

deterioration itself, cause more difficulties for the sufferer and their carer as they have to come to terms with the sufferer's change in personality, behaviour and character. In the later stages of HD full nursing care is required and secondary illnesses such as pneumonia are often the actual cause of death rather than the disease itself. Moreover, for every HD patient, it is reported that there are another 20 people (including those who are at risk) who suffer the consequences of HD (Hayden et al, 1980). This may be in relation to caregiving, genetic inheritance or the sheer burden that such a devastating disease places upon a family system.

1. The Clinical Characteristics of Huntington's Disease:

History.

Huntington's Disease (HD) has a long history that stretches back for well over a century. HD (then referred to as Huntington's Chorea (HC) was originally described by George Huntington in 1872. It was this description by Huntington (1872, cited in Harper, 1996) that defined the condition as separate from other forms of previously documented chorea.

Huntington's (1872) paper is mostly an overview of chorea in general. However, the final part describes for the first time the type of chorea that we know as HD today.

" ... And now I wish to draw your attention more particularly to a form of the disease which exists, so far as I know, almost exclusively on the east end of Long Island. It is peculiar in itself and seems to obey certain fixed laws. In the first place, let me remark that chorea, as it is commonly known to the profession, and a description of which I have already given, is of exceedingly rare occurrence there. I do not remember a single instance occurring in my father's practice, and I have often heard him say that it was a rare disease and seldom met with by him.

The hereditary chorea, as I shall call it, is confined to certain and fortunately a few families, and has been transmitted to them, as an heirloom from generations away back in the dim past. It is spoken of by those in whose veins the seeds of the disease are known to exist, with a kind of horror, and not at all alluded to except through dire necessity, when it is mentioned as 'that disorder'. It is attended generally by all the symptoms of

common chorea, only in an aggravated degree hardly ever manifesting itself until adult or middle life, and then coming on gradually but surely, increasing by degrees, and often occupying years in its development, until the hapless sufferer is a quivering wreck of his former self. It is as common and is indeed, I believe, more common among men than women, while I am not aware that season or complexion has any influence in the matter. There are three marked peculiarities in this disease: 1. Its hereditary nature. 2. A tendency to insanity and suicide. 3. Its manifesting itself as a grave disease only in adult life.

1. *Of its hereditary nature. When either or both the parents have shown manifestations of the disease, and more especially when these manifestations have been of a serious nature, one or more of the offspring almost invariably suffer from the disease, if they live to adult age. But if by any chance these children go through life without it, the thread is broken and the grandchildren and great-grandchildren of the original shakers may rest assured that they are free from the disease. This, you will perceive differs from the general laws of so-called hereditary diseases, as for instance in phthisis, or syphilis, when one generation may enjoy entire immunity from their dread ravages, and yet in another you find them cropping out in all their hideousness. Unstable and whimsical as the disease may be in other respects, in this it is firm, it never skips a generation to again manifest in another; once having yielded its claims, it never regains them. In all the families, or nearly all in which the choreic taint exists, the nervous temperament greatly preponderates, and in my grandfather's and father's experience, which conjointly cover a period of 78 years, nervous excitement in a marked degree almost invariably attends upon every disease these people may suffer from, although they may not when in health be over nervous.*
2. *The tendency to insanity, and sometimes that form of insanity which leads to suicide, is marked. I know of several instances of suicide of people suffering from this form of chorea; or who belonged to families in which the disease existed. As the disease progresses the mind becomes more or less*

impaired, in many amounting to insanity, while in others mind and body both gradually fail until death relieves them from their sufferings. At present I know of two married men, whose wives are living, and who are constantly making love to some young lady, not seeming to be aware that there is any impropriety in it. They are suffering from chorea to such an extent that they can hardly walk, and would be thought, by a stranger, to be intoxicated. They are men of about 50 years of age, but never let an opportunity to flirt with a girl go past unimproved. The effect is ridiculous in the extreme.

3. *Its third peculiarity is its coming on, at least as a grave disease, only in adult life. I do not know of a single case that has shown any marked signs of chorea before the age of thirty or forty years, while those who pass their fortieth year without symptoms of the disease are seldom attacked. It begins as an ordinary chorea might begin, by the irregular and spasmodic action of certain muscles, as of the face, arms etc. These movements gradually increase, when muscles hitherto unaffected take on the spasmodic action, until every muscle in the body becomes unaffected (excepting the involuntary ones), and the poor patient presents a spectacle which is anything but pleasing to witness. I have never known a recovery or amelioration of symptoms in this form of chorea; when it once begins it clings to the bitter end. No treatment seems to be of any avail, and indeed nowadays its end is so well-known to the sufferer and his friends that medical advice is seldom sought. It seems at least to be one of the incurables" (Huntington, 1872: 320-321).*

It can be seen that all of the fundamental features of HD as it is known today are identifiable within Huntington' s (1872) description i.e. the autosomal pattern of genetic inheritance, the adult onset, progressive disease course, and fatal outcome; choreic movement disorder combined with mental (cognitive) impairment, sexual inappropriateness and possible risk of suicide (psychiatric disorder).

Neurology.

Although Huntington's (1872) description does summarise the core clinical features of HD, it leads to the assumption of a reasonably straightforward and easily recognisable clinical presentation. However, over the past century, much research has been conducted that has identified a number of ways in which HD may present (Harper, 1996). The identification of a polymorphic DNA marker genetically linked to HD (Gusella et al, 1983) and subsequently the gene's pathogenic mutation (Huntington's Disease Collaborative Research Group, 1993) have further allowed clinicians to recognise more accurately the multiple manifestations of HD. Due to such research, diagnosis is no longer purely based on clinical features (and neurological confirmation at death). Genetic testing can be implemented in order to give a definitive prognosis during a patient's lifetime.

Onset of HD:

Although HD can present as early as the age of 2 (Huntington's Disease Collaborative Research Group, 1993), it is generally considered to be a late onset disease with a median onset age of mid-forties to early fifties with wide variation (Kremer, 2002). There is no single presenting sign or symptom in HD. In its earliest phases, there is an insidious and slow deterioration of intellectual function as well as mild personality changes. The clear appearance of extrapyramidal signs such as chorea, hypokinesea, rigidity or distonia mark a phase in the progression of the disease rather than the onset of the disease itself. Penny et al (1990) suggest that prior to these signs, most individuals will only display minor motor abnormalities such as general restlessness, abnormal eye movements, hyperreflexia, impaired

(piano like) finger tapping or rapid alternating hand movements which may increase during stress and mild dysarthria. Such minor abnormalities can precede more obvious signs of extrapyramidal dysfunction by three years or more (Snowden et al, 1998; Kirkwood et al, 2000).

Mid-course disease.

The middle stages of HD bring about motor disorder, which is probably the feature that is classically associated with the disease. This stage is subjugated by visibly motor abnormalities including extrapyramidal (i.e. outside of the pyramidal tracts) signs and more general non-specific impairments of essential skilled movements related to dysphagia (swallowing difficulties), dysarthria (speech disturbances) and gait. Therefore, there are two parts of the movement disorder, the presence of involuntary movements and the impairment of voluntary movements.

Involuntary (extrapyramidal) motor abnormalities:

Chorea is the major motor sign of HD and has been defined as *"a state of excessive, spontaneous movements, irregularly timed, randomly distributed and abrupt. Severity may vary from restlessness with mild, intermittent exaggeration of gesture and expression, fidgeting movements of the hands, unstable dance-like gait to a continuous flow of disabling, violent movements"*. (Lakke, 1981: 314).

Choreic movements are continuously present during the waking hours of an HD affected individual, they cannot be voluntary suppressed by the patient and are worsened by stress,

anxiety and depression (Rosenblatt et al, 1999). Chorea is a feature of HD in over 90 percent of patients (Kremer, 2002). As the disease progresses, chorea tends to disappear and is replaced by bradykinesia (abnormal slowness of movement) and rigidity (Young et al, 1986), hypokinesia (Van Vugt et al, 1996) and dystonia (Young et al, 1996).

Other (voluntary) motor abnormalities.

Oculomotor disturbances are present in the vast majority of HD affected patients (Lasker & Zee, 1997). Early on, the patient may lose the inability to suppress reflexive glances to suddenly appearing novel stimuli and may also have a delayed initiation of voluntary saccades (Lasker & Zee, 1997). Later in the disease process, slowing of saccades is seen in up to 75% of patients and they may also suffer from, impairment of gaze fixation, slowing of the optokinetic nystagmus and ability to suppress blinking (Penny et al, 1990). Patients may also suffer from the impairment of voluntary motor functions causing clumsiness in common everyday activities. Disturbances can be found in motor speed, fine motor control and gait (Thompson et al, 1988). Such symptoms appear to be correlated with disease progression and as such are a better measure of duration of illness than chorea (Folstein et al, 1983).

End-stage disease.

In the final stages of HD, most patients will experience a loss of independence, severe restrictions in functioning and dependence upon others for their daily living. In these final

stages of the disease, full nursing care is required (Kremer, 2002) and secondary illnesses such as pneumonia, choking, nutritional deficiencies and skin ulcers are often the actual cause of death rather than the disease itself (Lanska, et. al, 1998). The advanced stages of HD are dominated by hypokinesia, bradykinesia, rigidity and dystonia although choreic movements may still be visible. Severely impaired speech or mutism may impair communication to a large extent. It is during this phase that the patient may lose access to some of the experiences that have been important to him/her during their lifetime through the inability to communicate. For example, if the patient is unable to communicate effectively what he/she likes to eat, wants to watch on the television, what clothes are comfortable etc, their quality of life may be diminished especially in cases where there are no family members present to fill in these gaps of information (Parker et al, 2002). Swallowing is also impaired and patients may be fitted with a percutaneous endoscopic gastronomy (PEG) tube in order to ensure that they receive their required daily intake of calories and to further reduce the possibility of choking (Rosenblatt et al, 1999).

Patients also lose their ability to walk as their gait becomes further disturbed and will ultimately be confined to either bed or a wheelchair. Furthermore, a combination of muscle hypertonia and increased tendon reflexes can be indicative of upper motor neurone dysfunction. Unfortunately, a large number of patients are also on psychotropic medication such as benzodiazepine sedative, antidepressants or neuroleptics prescribed as antichoreic drugs which can also exacerbate a problematic gait (Rosenblatt et al, 1999).

Weight loss also features as a symptom of late stage HD and may even occur in conjunction with increased dietary intake (Farrer & Yu, 1985) although swallowing is suggested as being a

major course of such emaciation (Kremer, 2002). Furthermore, sleep is often disturbed in the advanced stages of the disease (Silvestri et al, 1995).

Duration of HD.

The median duration of HD is between 15 and 20 years with no effect of sex (Roos et al, 1993; Foroud et al, 1999) However, there is marked individual variation with the disease course being as long as 45 years in some instances. One of the factors that may influence a longer disease course / survival rate is the CAG repeat size with age at death and repeat size being significantly correlated (Andrew et al, 1993).

Neuropsychology and Neuropsychiatry.

HD is characterised by movement abnormalities (i.e. chorea and dystonia), cognitive deterioration and affective disturbances (Folstein, 1989). Therefore it spans a triad of motor, cognitive and psychiatric symptomology. Although the motor symptoms are the most clearly visible, there is much evidence to suggest that the non-motor symptoms have the biggest impact on the patient's life.

Cognitive Changes.

The cognitive features of HD are present early on in the course of HD and become more severe as the disease progresses (Brandt & Butters, 1996). However, there is wide variation

in how they present. In some affected individuals such impairments are obviously recognisable to family members and evident during clinical interviews. However, in other individuals the changes are more subtle and may only be detected at a neuropsychological examination.

Dementia.

The dementia of HD includes a deficit in the metamemorial control processes that orchestrate retrieval efforts (i.e. executive functions thought to be dependent on the prefrontal cortex and its striatal connections). Therefore, patients present with a flat retrograde amnesia profile, impairment in the acquisition of perceptual skills thought to be a feature of cognitive loss and impaired biasing performance which reflects a deficit in the development of central motor programmes. There is also evidence of the dependence of central motor programmes on basal ganglia structures and impairment on explicit memory tasks involving the learning of skill based knowledge (Brandt, 1991).

Features of early dementia syndrome often coincide with onset of motor signs. In the early stages of disease problems with aspects of attention and memory (especially procedural memory), visuomotor and visuographic skill, and executive functions are evident as well as primary sensory and perceptual abilities, most aspects of language, nonmotor spatial cognition and recognition memory (Rothlind et al, 1993).

Attention-demanding cognitive operations are among the first to deteriorate in HD and there are only a few psychometric tests that assess aspects of attention in isolation.

Attentional difficulties however, can be inferred from poor performance on traditional tests such as the WAIS (Nelson, 1976) and Stroop (Stroop, 1935). A more specific scale, The Unified Huntington's Disease Rating Scale (UHDRS; Huntington's Disease Study group, 1996) is a rating system that has been developed to quantify the severity of all aspects of Huntington's Disease. It is divided into multiple subsections: motor, cognitive, behavioural, functional and aims to provide a uniform assessment of the clinical features and course of HD. The UHDRS has undergone extensive reliability and validity testing (Huntington's Disease Study Group, 1996) and has been used by them as a major outcome measure in controlled clinical trials. However, it is relatively expensive to obtain and not available to researchers or clinicians to use without the use of a training tape. This may make it inaccessible to many carer groups or nursing homes who could benefit from its application.

Language.

Despite the progressive nature of HD, clinically significant aphasia is rarely seen. However, motor speech impairments such as dysarthria (imperfect speech articulation) affect 50% of early stage patients (Brandt & Butters, 1996). Speech disorders become more pronounced as the disease progresses, and patients may be unable to communicate intelligibly in the end stages of the disease. Performance on language tasks (as opposed to speech itself) remains normal although HD patients may initiate verbal communication less often and participate very little in ongoing conversation. Patients tend also to have long response latencies to questions and pronounced intervals between phrases (Podoll et al, 1988). As such, conversation is interspersed with long gaps of silence and has reduced syntactic complexity (Folstein, & McHugh, 1983). In advanced HD, spoken language consists of single words or

short phrases that often do not constitute complete sentences. In contrast to this marked reduction in complexity, syntactic structure remains correct and speech content is usually very appropriate until advanced illness (Gordon & Illes, 1987). Patients have marked deficits in retrieval but little breakdown in semantic knowledge (Martin & Fedio, 1983), although in later stages, some mild deterioration of semantic knowledge may occur. Such specific deterioration means that patients who cannot communicate effectively with the outside world may still have a good understanding of what is happening around them.

Spatial skills and perception.

Deficits in visuomotor performance are evident even in mild HD patients (e.g. Bamford et al, 1989), although true constructional apraxia is rarely noted (Brandt & Butters, 1996). The ability to cope with even simple geometric designs is impaired in early stage HD (Mohr et al, 1997). Such deficits are thought to be due to striatal pathology or damage to frontal striatal pathways (Potegal, 1971).

Memory.

Memory disturbances are very prominent and present as an early cognitive feature of HD (Moses et al, 1981). Deficits are displayed in the learning and retention (Moses et al, 1981) and retrieval of information (Brandt, 1985). Butters et al. (1990) argue that in addition to problems with explicit memory, there is considerable evidence that specific forms of implicit memory are also impaired in HD.

Executive functioning.

Early stage patients describe problems with planning, organising and scheduling day to day activities (Watkins et al, 2000). Patients become less adaptable and behaviourally rigid, getting stuck in an idea or task (Pillon et al, 1991). In early HD, impairment of daily functioning is more likely to result from these cognitive deficits than from motor impairments. Deficits are found in impaired attention, decreased verbal fluency, poor motor programming, difficulty compensating for postural adjustments, inability to switch off cognitive sets and difficulties with abstraction (Josiassesn et al, 1983; Alexander et al., 1989).

Non - cognitive features of HD.

Depression.

In addition to the movement disorder and dementia, patients often present with prominent affective disturbances which have been referenced widely in the extant literature (e.g. Huntington, 1872; Heathfield, 1967; Bolt, 1970; Folstien et al, 1983, 1987). Patients with major depression have a sustained low mood, feelings of worthlessness and guilt, apathy, loss of energy, loss of appetite and changes in sleep patterns. In severe cases, patients may also experience delusions or hallucinations. Depression in HD can be difficult to diagnose as patients may not complain about a low mood, and symptoms may be masked by other clinical features of HD. For example, loss of appetite in depression may be masked by an increase in appetite often seen in HD (Craufurd, & Snowdon, 2002).

Suicide.

The potential for death by suicide in HD has been recognised since Huntington's first description (Huntington, 1872) and also more recently (e.g. Hayden et al, 1980; Schoenfield et al, 1984). In an American study investigating this, the proportion of death due to suicide among persons with HD is almost 4 times greater than the corresponding proportion for the U.S. Caucasian population (Farrer 1986). It still uncertain whether suicide may in fact be a rational (albeit) extreme response to an intolerable inherited situation or a manifestation of dementia (Hayden, 1981). High rates of suicide have also been linked to major depression (Wood et al, 2002). In such cases suicidal patients can be treated effectively with antidepressant drugs (Rosenblatt et al, 1999). .

Mania.

Whilst depression is the most common psychiatric problem found in HD, a small number of patients become manic, displaying elevated or irritable mood, overactivity, decreased need for sleep, impulsiveness and grandiosity. Healthfield (1967) described four patients out of eighty with hypomania and delusions of grandeur and Bolt (1970) noted grandiose ideas in eleven out of three hundred and thirty four cases. Folstein et al (1987) found hypomania in approximately 10% of their patients.

Schizophrenia-Like disorder

There are many reports of schizophrenic-like symptoms in HD. Guttermann (1938) describe one patient with frank schizophrenic illness and Heathfield (1967) reported six cases of paranoid schizophrenia. Such diagnosis is difficult in HD as symptoms such as emotional withdrawal and emotional blunting are found in both conditions. In some cases, the onset of schizophrenic-like symptoms may precede the onset of motor symptoms and patients may be misdiagnosed as suffering from schizophrenia (Rosenblatt et al, 1999).

Irritability.

Irritability and bad-tempered outbursts are one of the most common and troublesome behaviours associated with HD. Irritability is often associated with a depressed mood, but can also result from a loss of the ability of the brain to regulate the experience and expression of emotion (Rosenblat et al, 1999). Often family members will complain that the patient has become irritable for no obvious reason and that even the slightest provocation at this time may provoke an outburst of angry or violent behaviour (Craufurd & Snowden, 2002). Heathfield (1967) describes 15 out of 80 patients studied as having aggression and irritability and Bolt (1970) reported 50% of three hundred and thirty four cases has some 'degree of ill-humor' in the form of either irritability, aggressiveness or rage. Such irritability may be a side effect of drug therapy and can also be a sign of depressive illness (Harper, 1996).

Apathy.

Patients with HD may also present with a visible loss of motivation, initiative and spontaneous expression (collectively termed as 'situational apathy') which is particularly common in the middle and later stages of the disease (Caine and Shoulson, 1983). Apathetic patients become unmotivated and uninterested in their surroundings and lose enthusiasm or spontaneity (Rosenblat, et al, 1999). Burns et al (1990) found apathetic behaviour to be present in 48 percent of HD patients they studied. Oliver (1970) suggested that apathy may be a secondary symptom of depression. However this has been challenged by Mayberg et al (1992) who found that most patients suffer from apathy in the later stages of HD whether or not they present with affective disorder.

Sexual Disorder.

Despite Huntington's (1872) descriptions of sexual deviation in HD, it is more often the case that patients become uninterested in sexual activity (Craufurd et al, 2001). However, Dewhurst et al's (1970) paper is often cited as evidence of frequent hypersexuality in HD patients. Fedoroff et al (1994) studies sexual dysfunction in a sample of 39 HD patients and noted that although sexual disorders are common, it is evident that hyposexuality is far more common than hypersexuality with 62 percent of patients reporting a loss of libido (confirmed by their partners) and only 6 percent reporting sexually disinhibiting (such as public masturbation or voyeurism) behaviours.

Summary.

Cognitive and psychiatric changes are an integral aspect of HD as they greatly affect the patients' functional capacity. HD patients experience cognitive changes that are bound to place limitations on their capabilities. However, psychiatric changes can often be treated effectively once diagnosed. It is probably the behavioural changes that are the most difficult aspect of HD for both the patient and the carer as they come to terms with the sufferer's change in personality, behaviour and character (Harper, 1996).

2. The Genetics of Huntington's Disease:

Basic Genetics.

HD is a genetic condition inherited as an autosomal dominant trait with complete life time penetrance. Each child of an affected person has a 50% chance of inheriting the gene. All gene carriers will inherit the illness if they live long enough. The gene and mutation for HD was only identified in 1993 (Huntington's Disease Collaborative Research Group). However, Huntington (1872) noted in his earliest description that HD was a dominant hereditary disease: "... .*One or more of the offspring almost invariably suffer from the disease if they live to adult age. But if by any chance these children go through life without it, the thread is broken and the grandchildren and great-grandchildren of the original shakers may rest assured that they are free from the disease.*" (Huntington, 1872 , pg. 321).

The genetic locus for HD has been localised to the short arm of chromosome 4 (Guzella, 1983). This gene mutation consists of the expansion of a region in which there is a sequence of the 3 nucleotides cytosine, adenine and guanine (CAG) that normally repeats between 11 and 34 times. On HD chromosomes the region expands over 37 times. This expansion of the gene which produces a protein called Huntingtin, damages the nerve cells in the basal ganglia and cerebral cortex (Aylward et al, 1997). The degree of expansion from the HD parent to child can vary but appears to be greater with paternal than maternal transmission (Duayo et al, 1993). A greatly expanded gene is associated with early onset of illness (e,g, Snell et al, 1993) as well as rapid progression (Brandt et al, 1996). Furthermore, CAG repeat

length seems to account for 47-73 percent of the variation in age of onset (Rosenblat et al, 2001).

Epidemiology.

Because of the genetic and therefore kindred nature of HD, most of the information available with regards to the genetic nature of HD is by means of family surveys. HD is a universal disease although there do appear to be some particular 'hot spots' around the world possibly due to founding ancestors and lack of emigration (Penney et al, 1990). Folstein et al (1987) further report some phenotypic variation among racial groups.

Early studies in North America suggested a common ancestry from a small number of founding immigrants, and it was initially thought that the gene has come principally from the original migrants from East Anglia, England (Jelliffe, 1908; Davenport and Muncy, 1916). However, this was found to be inaccurate (Caro, 1977) and a broader 'northern European' origin for the gene was accepted.

HD has been observed in the USA, Canada, South America, Britain and the majority of Europe, Asia, Australia, The Pacific Islands, Japan, Africa, South Africa and Mauritius (Bates et al, 2002). Its prevalence ranges from 0.37 per 100, 000 in Hong Kong (Leung et al, 1992) to 17.4 per 100,000 in Tasmania (Conneally, 1984). Estimates of universal prevalence range from 5-8 per 100,000. However, if this is restricted to the population between the ages of 40 and 55, prevalence rises to 12 per 100,000 (Harper, 1991). In the UK, prevalence was

estimated in 1991 to be 6.4 per 100,000 (region of Northern Island) (Morrison et al, 1995). Although HD is evident across the UK, it is particularly prevalent in North East Scotland (9.95 per 100,000; Simpson and Johnston, 1989) and South Wales (8.85 per 100, 000; Quarrell et al, 1988).

Presymptomatic testing.

The discovery of the genetic marker that localised the gene for HD to the short arm of chromosome 4 (Gusella, 1984) paved the way for presymptomatic testing in HD for individuals at risk of having the HD gene. The test has provided the opportunity for individuals to establish not only whether they carry the gene so they can plan for the future but also, can be used in prenatal testing, giving parents the opportunity to terminate a pregnancy if the feotus is found to carry the gene. Testing embryos prior to termination has also become possible thus, giving couples the opportunity to have a pregnancy without prenatal testing.

However, the availability of a presymptomatic test also raises concerns in terms of psychological morbidity. Farrer (1986) identified that between 11 and 33 percent of people 'at-risk' from HD have considered suicide as a possibility. Therefore, if such individual's fears are confirmed, there may be a possible increase in death by suicide amongst identified carriers (Tibben 2002). Farrer (1987), Kessler (1987), Brandt et al, (1989) and Huggins et al (1992) have made reference to the potential for further problems such as depression, family turmoil, divorce and survivor guilt. Moreover, the medical value of presymptomatic testing

is unclear as there is no way at present to control the onset of the disease process and no effective cure or treatment (Martin, 1984). In more recent years, employment and insurance discrimination have also been evident amongst those who are found to carry the gene (Pincus, 2001).

As well as impacting upon the individual, presymptomatic testing also has implications for other family members. Hans and Koeppen (1989) noted that partners may often react with disbelief and denial. However, this initial reaction may turn to resentment and hostility as they become aware of the possibility of transmission to their children (see also Kessler, 1988; Kessler & Bloch, 1989; Codori & Brandt, 1994; Sobel & Brookes Cowan, 2000; Williams et al, 2000). Because of the hereditary nature of the disease, presymptomatic testing has far reaching consequences within families. For example, adult children may want to be tested even if 'at-risk' parents have not been tested. As such, a positive test result would be evidence of the parent(s) positive status. Or, as this following example illustrates, sometimes the issue for prenatal testing comes before the 'at-risk' individual is ready to know their genetic status.

Example (taken from Smith et al, 1998, p 42): Kirsten is a twenty-nine-year-old woman whose husband, David, is at 50 percent risk for HD. She contacts a predictive testing center, reporting that she has discovered she is six weeks pregnant and wants to have the fetus tested for the HD mutation. In a counseling session, Kirsten and David reveal that the pregnancy in unplanned and that David is undecided about whether he wants the predictive test for himself. Kirsten, however, feels strongly that she does not want their child to be at any risk for HD. Now that the technology is available, she wants to use it to enable her to decide whether to continue the pregnancy.

Kirsten is aware that a positive prenatal result would reveal that David has the HD mutation.

Nondisclosing prenatal exclusion testing is not an option, as both of David's parents are deceased.

Kirsten insists that she be allowed to proceed with prenatal testing. David however, does not feel prepared emotionally for the possibility of receiving two positive tests results at once and then proceeding with a termination of the pregnancy. He believes they should continue the pregnancy. He believes that if the baby is a gene carrier, preventative treatment will be available by the time he or she is an adult.

This particular example highlights the issues faced both by 'at-risk' individuals and their families. If the test is taken, 'David' does not feel able to cope with the situation. However, it is 'Kirsten' who will be left with the burden of caring for both a husband and a child if the test is not taken and they are eventually both found to be carriers. Thus, the decision not to test could potentially place 'Kirsten' in a caregiving role for many decades.

Therefore, genetic testing will inevitably have a profound effect on the family. Spouses particularly have difficulty coming to terms with test results regardless of whether they are positive or negative (e.g. Hans and Koeppen, 1980; Evers-Kiebooms et al, 1990; Kessler, 1993; Tibben et al, 1992, 1993; Quaid & Wesson, 1995; Codori & Brandt, 1994).

Summary.

HD is a genetic condition that is passed through families as an autosomal dominant trait. As such, any offspring of an HD affected parent have a 50% chance of inheriting the gene for HD. HD has complete life time penetrance meaning that if an individual has the gene for HD and they live long enough, they will get the disease. Complications arise with HD as it generally presents in middle adulthood, a time when children may have already been born. Genetic testing for HD has meant that individuals over the age of 18 can establish their

genetic status before symptoms arise, and as such make life choices with regards to children. However, there are concerns with regards to the ethics of testing for a disease for which there is no cure.

Chapter 3.

Quality of Life.

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Introduction.

Since the 1960's, 'quality of life' (QoL) has been emerging as a useful outcome measure by which to judge the efficacy of psychological interventions (Cummins, 1997; Land, 2000; Rapley, 2003). Previous to this, health outcome indicators (defined as indicators of change in health status) traditionally included information on avoidable mortality, survival rates, symptom relief, pain and physical and biomedical markers of recovery (Donabedian, 1985). There are many documented concerns about the conceptualisation of QoL (e.g. Cummins et al, 1994; Allison, Locker & Feine, 1997; Anderson & Burckhardt, 1999. However, such concerns can often be addressed through the use of a well- operationalised and validated tool (e.g. Cummins 1997; The WHOQOL group, 1998). Using QoL as a measure of outcome focuses on the impact of a condition or situation on the *individual's* emotional and physical functioning and lifestyle. Therefore, QoL indicators have an added advantage of comparing the benefits of different interventions and further provide means of quantifying risk / benefit ratios and assessing the subjective benefit of interventions (Rabins, 2000). As such, they can help to answer the question of whether an intervention leads to an increase in wellbeing by providing a more client-led baseline against which the effects of the intervention can be evaluated (Bowling, 2001).

1. Defining Quality of Life

Brief History.

In Western tradition, discourse on the concept of 'quality of life' (QoL) dates back to Aristotle (384-322BC) and early Greek Philosophy (see Veenhoven, 1991). Although the term QoL was not in existence at this time, Aristotle made two very important points. Firstly that QoL means different things to different people and secondly that it was context specific. The term 'quality of life' was rarely mentioned until the 20th century (Fayers & Machin, 2001) and the notion of quality of life in relation to health followed on from this. One of the earliest statements of QoL in relation to health comes from The World Health Organisation's (WHO, 1948) definition of QoL that noted three domains of physical, mental and social wellbeing within the context of disease.

The earliest attempts to assess patients from a non-biological perspective appear to have been objective measurements of functional health status i.e. the ability to perform routine self-care and complete basic physical activities and a level of independent living. One of the first attempts to measure life quality in terms of functional ability was the New York Heart Association Classification (1939, cited in Fayers & Machin, 2001) which evaluated the functional capacity of patients with Heart Disease. In 1948, Karnofsky proposed a performance scale for use in clinical settings. This was a single numerical scale that gave scores from 0 (for dead) to 100 (no evidence of disease) for a combination of three factors: the ability to carry out normal activities, the need for custodial care and the need for medical care. Over the following thirty years, other scales were developed to measure functional

deterioration (The index of Independence of Activities of Daily Living, Katz et al, 1958, 1970), rehabilitation potential (The Barthel Index, Mahoney & Barthel, 1965) and physical functioning (Instrumental Activities of Daily Living Scale, Lawton & Brody, 1969). Simultaneously, a number of social science indexes were proving to be useful to sociologists and psychologists as social indicators of health. After the WHO has defined health as *"not only the absence of infirmity and disease, but also a state of complete physical, mental and social well being" (WHO, 1948),* health care professionals were reminded that the patients' health was more than just a physical state and could be affected by both environmental and social factors as well.

In the early 1960's, the Social Indicators movement led by psychologists and sociologists, began to advocate a broader assessment of life quality by assessing changes such as education, health, employment, crime victimization, political participation and population growth. These notions were made available through a collection of essays that referred to measuring various aspects of society and comparing them to the ideals of the nation (Gross, 1966). Three years later, the Department of Health, Education and Welfare published 'Towards a Social Report' (1969) in which they noted that although there were numerous measures of death and illness, there were no measures of physical vigor or mental health. They further noted that although there were statistics on the level and distribution of income, there were no measures of the satisfaction that such income brought. By 1972, there were more than 1000 articles that related to Social Indicators (Wilcox et al, 1972) and articles on "quality of life" in relation to philosophical concepts, design and testing of new instruments and studies using those instruments began to emerge.

The first quality of life scale to become popular was Priestman & Baum's (1976) adaptation of linear analogue self-assessment (LASA) methods (i.e. visual analogue scales, VAS) to measure QoL in breast cancer patients. On a ten-centimeter line labelled with extreme anchors at each end, patients would place a mark corresponding to their feelings at the moment. The ten questions in the index ranged from feelings of well being, to pain, to the patient's perception of the treatment they were receiving. The sum of the marks became the overall measure of quality of life for that person (see also Andrews & Withey, 1976; Bradburn, 1969 for early subjective QoL measurements). However, by the late 1970s and early 1980s objective measurements were being developed to assess general health status such as the Sickness Impact Profile (Bergner et al. 1981) and the Nottingham Health Profile (Hunt, McEwan & McKenna, 1985). The Sickness Impact Profile is a behaviorally based measure of health status, which demonstrates good convergent validity, internal consistency and test-retest reliability. This instrument is frequently described (and used) today as a QoL questionnaire although it was not originally designed to measure the concept of QoL (Fayers & Machin, 2001). The Nottingham Health Profile was developed as a generic health-related quality of life measure and is used to assess physical functioning, physical and psychological symptoms, impact of illness, perceived distress and life satisfaction. It is also well used in research today and demonstrates good validity, internal validity and test-retest reliability (Hunt et al 1985, Jenkinson, et al, 1988, Bowling, 2001).

In 1985, the Food and Drug Administration's (FDA) decision to require QoL data as one of the 'Key efficacy parameters' in a clinical trial for new anti-cancer agents paved the way for QoL measurements to be used as outcome indicators in clinical trials. The FDA reported it

would be willing to approve a drug in certain cases only if it reduced pain or toxic effects. A working group from the National Cancer Institute (1991) and the FDA later recommended that validated QoL measures would be useful for comparing either pre and post treatment groups or treatment versus placebo groups. In addition, QoL assessment was used as the primary outcome in a randomized trial to examine the QoL of patients taking one of three anti-hypertensive medications (Croog et al, 1986). Satisfaction with life, physical state, emotional state, intellectual state, social functioning and wellbeing were all assessed, allowing the pharmaceutical manufacturers to promote their products for not only their biomedical effect but also their positive effect on QoL.

Much of the development of QoL instruments has been built on early scales with newer instruments placing more emphasis on subjective indicators of QoL such as emotional, role, social and cognitive functioning. Therefore, the concept and measurement of QoL as we know it today is derived from both indexes of health status and indexes of happiness, well-being and other 'affects'. In the current research climate, a sharp increase in QoL papers and measures is evident. The World Health Organisation (WHO) alone have hundreds of publications in quality of life research and have extensively developed a number of tools that have been used in a variety of different populations. This has now grown so extensively that no one researcher could thoroughly review all of the current literature and it is the sole focus of the international journal, *Quality of Life Research*. However, despite this growth, there are a number of concerns and criticisms of QoL tools and their usefulness in assessing life quality, which will be addressed in the next section.

Conceptualisation.

The World Health Organisation (WHO) define Quality of Life as, *"... .an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns"*. (WHO Group, 1995, pg 3). This is a broad ranging concept which incorporates the individual's physical health, psychological state, level of independence, social relationships, personal beliefs and relationship to salient features of the environment. Furthermore, this definition highlights the view that quality of life refers to a subjective evaluation, which includes both positive and negative dimensions, and which is embedded in a cultural, social and environmental context.

However, there is no agreed definition or standard form of measurement of QoL and as such, the QoL construct has become very complex in composition. There are many documented concerns about the conceptualisation of QoL (e.g. Cummins et al, 1994; Allison, Locker & Feine, 1997; Anderson & Burckhardt, 1999). Cummins (1996) has recorded well over 100 instruments which purport to measure life quality in some form, but each one contains an idiosyncratic mixture of dependent variables. Moreover, there is little empirical research attempting to define those qualities which make life and survival valuable. The literature covers a range of components from which QoL are often derived, such as functional ability, including role functioning (e.g. domestic, return to work), the degree and quality of social and community interaction, psychological well-being, somatic sensation (e.g. pain) and life satisfaction. Therefore, QoL is recognised as a concept representing individual responses to the physical, mental and social effects of illness on daily living which influence the extent to which personal satisfaction with life circumstances can be achieved. Moreover,

in recent years quality of life has become the driving force in service design, delivery and outcome evaluation, from medicine to social care (Rapley, 2003). Quality of life is regularly used to justify money well spent as it has become equated to satisfaction with a service or outcome and therefore, value for money. However, issues in terms of operationlisation of the construct and whether to measure it objectively or subjectively make questionable the value of QoL measures and their generalisability and application, both within and between different populations.

Rapley (2003) notes that much QoL research is predicted upon a priori acceptance of the notion of QoL as a hypothetical construct which through objective criteria can be measured and quantified. Such conjecture underpins the development of all QoL scales which attempt to quantify the quality of an individual's or a communities life through a meaningful yet numerical description. However, there has been much criticism of the notion of objective quality of life (Cummins, 1998) with researchers reporting differences between objective measures and subjective interpretation of QoL (Bowling & Windsor, 2001) and current research leaning towards the use of subjective measures of perceived well-being in order to attempt to quantify life quality. Subjective measures allow for individual differences to dictate the relevance and importance of any given situation and therefore its impact on perceived quality of life. However, there are clearly concerns with measures of subjective well-being ultimately being used to assess service quality (Hatton & Ager, 2002) or even as evidence of the effectiveness of therapeutic interventions as one's subjective interpretation of a situation may differ vastly from its 'objective reality'. Thus leaving the question open as to whether the objective situation or perceived issues are the real areas of concern for health care professionals, patients and researchers alike.

Summary.

Definitional diversity seems to be one of the key problems in conceptualizing QoL (Rapley, 2003). As such, QoL research is often seen as poor and questionable as to whether it actually measures what it purports to. Meeberg (1993) argues that the term is frequently overused and underdefined and the definition of quality of life is still imprecise (Felce, 1997). Therefore, although it is widely agreed that QoL is indeed a multi-dimensional construct, there is still no consensus as to the number or variety of dimensions that should be used within QoL research (Rapley, 2003) or indeed an ultimate definition of what QoL really is.

2. Researching Quality of Life within Health Psychology.

Using Quality of life as an outcome measurement.

Bowling (2001) notes that in the current day, purchasers of health care are generally expected to allocate resources on the basis that such resources are indeed advantageous and effective. As such, physical and therapeutic interventions need to have some proved 'health gain' in order for them to be seen as effective and beneficial to the individual. However, traditional objective outcome indicators such as physical functioning (e.g. The Barthel Index, 1965), do not take into account the subjective impact of a situation on the individual. Using QoL as a measure of outcome, though, has the benefit of focussing on the impact of a condition or situation on the *individual's* emotional and physical functioning and lifestyle. Therefore, QoL indicators have an added advantage of comparing the benefits of different interventions and further provide means of quantifying risk / benefit ratios and assessing the subjective benefit of interventions (Rabins, 2000). As such, they can help to answer the question of whether an intervention leads to an increase in wellbeing as they provide a more individual-led baseline against which the effects of the intervention can be evaluated.

QoL has therefore become more relevant to areas of Clinical Medicine and Health Psychology as an alternative or supplement to more objective clinical indicators of health. As such, the term 'Health Related Quality of Life' (HRQoL) is now often used to identify the dimensions of life quality that clinicians and outcome researchers are interested in. In general, HRQoL dimensions include general health, physical functioning, physical symptoms and toxicity, emotional functioning, cognitive functioning, role functioning, social wellbeing

and functioning, sexual functioning and existential issues (Fayers & Machin, 2001). However, there are concerns about the use of the term 'HRQoL' partly because it is a very ill defined term and studies vary as to which aspects of HRQoL should actually be assessed. As such, the term HRQoL still requires definition / operationalisation by the researchers of each individual study. Cummins (1998) disagrees with the notion of HRQoL, arguing that QoL should be a global construct that can be used to consider the wellbeing of larger populations rather than disease-specific sub-populations and individuals. Furthermore, researchers often use the terms HRQoL and QoL interchangeably making the revised terminology somewhat redundant. Despite such issues, measuring the health outcome of any intervention has become intrinsic to health service research (Leape et al, 1990).

Outcome Interventions in Caregiving.

There are a number of caregiver interventions that have proven to be effective in the alleviation of dementia caregiver distress and as such, help to improve QoL. For example, Hepburn et al (2001) report the benefits of dementia family caregiver training built on models of stress and coping. Furthermore, Hosaka and Sugiyama (2003) have provided physiological evidence of the benefits of therapeutic interventions. Hosaka and Sugiyama investigated the effects of a structured intervention programme which included education intervention, relaxation exercises and problem solving techniques on twenty Alzhiemer's dementia caregivers. They found that the interventions not only improved emotional and physical discomfort, but also improved immune functioning. However the benefit of interventions and their efficacy does appear to vary from study to study. Brodaty et al (2003)

conducted a meta-analysis of interventions for caregivers of individuals with dementia and noted significant benefits of interventions that were used to reduce caregiver psychological distress and increase caregiver knowledge. However they did not find any clear effect of interventions that were put into place in order to alleviate caregiver burden. Carradice et al (2003) also note a difference in the reported benefit of dementia caregiver stress interventions with subjective reports providing stronger evidence for the efficacy of interventions than objective measures. They argue that methodological rigor may 'dilute' any potential benefits of carer interventions and call for the use of theory driven interventions and research designs in order to reduce the inconsistencies between intervention studies.

Researching specific populations.

There is also evidence to suggest that specific caregiver populations may require specially adapted QoL interventions in order for them to have the most effect. Coon et al (2003) argue that no single, easily implemented and consistently effective method is available for achieving the same clinically significant outcomes across caregivers. As such, different populations of caregivers may require different educational interventions (Burgio et al, 2003). This appears to be especially important when considering racial and ethnic diversity (Coon et al, 2002).

Issues in terms of measuring interventions in relation to specific populations are further complicated in quality of life research by debates surrounding the utility of disease-specific or population specific QoL measures. Disease or population specific scales have the aim of

being able to both discriminate more finely between individual levels of life quality and further, of being more sensitive to clinical outcomes (Bowling, 2001). Researchers such as Morris (1990, cited in Bowling, 2001) have shown the benefit of using disease specific scales in identifying small but significant changes in health status and levels of disease severity. However, Kantz et al (1992) demonstrated the inability of a disease-specific quality of life measure to distinguish between treated and untreated patients. As such, disease-specific scales are often criticized for being too narrow in focus whilst neglecting to measure more general outcome modifying variables. The issues of global vs disease and population specific definitions and measures of QoL are closely related to the objective vs subjective QoL debate (Rapley, 2003). That is, is QoL measurement useful in terms of our overall understanding of what makes life meaningful for the population, or are we more concerned with highlighting issues that are pertinent to specific populations and subgroups. In order to develop solid theories and methods, general measures have the benefit of allowing researchers to consistently measure between populations. However, QoL issues that are intrinsic to certain populations with regards to disease, culture or other demographics may well not be 'tapped into' by such measures. As such, disease and population specific measures of QoL clearly have their place within both clinical interventions and psychological research and should perhaps be used in conjunction with more general measurements of quality of life, so as to allow consistency and comparison between different sub-groups of the population.

Summary.

Quality of life is a useful construct to use in outcome intervention research. However, there

are still many methodological and conceptual issues that are being debated with regards to the operationalisation of QoL, its objective and subjective components and further, the use of disease and population specific scales vs more generic determinants of QoL. Concerns with regards to defining QoL can to some degree be addressed through the use of well-operationalised research studies and validated tools. Moreover, it is generally accepted that there is a need for both objective measures of QoL that assess particular tangible needs and subjective indicators in order to allow the clinician or researcher to gain understanding of the impact of a situation on the *individual*. Thus, setting support in place though a client-led understanding of any given situation. Finally, the use of disease and population-specific measures of QoL can be a useful and beneficial supplement to more generic QoL measures that may not be sensitive to QoL issues that surround certain illnesses or situations. By combining disease and population specific measures with more generic QoL assessment tools, disease or condition-related attributes can be assessed whilst findings can still be generalised to other populations.

Chapter 4.

The impact of Huntington's Disease on the Quality of life of Spousal caregivers.

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Introduction.

Huntington's Disease (HD) is a chronic progressive dementia of the brain that causes movement abnormalities, cognitive deterioration and affective disturbances (Folstein, 1989). There is no cure for HD, with the treatments available being purely palliative or experimental. Furthermore, HD is a genetic condition inherited as an autosomal dominant trait with complete lifetime penetrance. In HD, the immediate family usually take on the responsibility of caring for an affected individual and often, the primary carer is the spouse (Kessler, 1993). Although there is a wealth of literature investigating the role of dementia family carers (Maslach, 1981), the symptoms and genetic nature of HD makes this carer role distinct from general dementia caregiving (Semple, 1995) and this can have implications for the professionals involved and also HD families. There is currently no work that explicitly investigates the impact of HD on the QoL of family carers, although the available literature would suggest that life quality is diminished for this carer group in terms of burden (Hans & Koeppen, 1980), gaining access to specific services (e.g. specialist aids and equipment; specialist advocacy and support) and dealing with professionals who are not always trained to deal with the family dynamics of HD.

Family Caregiving in HD.

It is generally the immediate family that takes on the responsibility of caring for an affected individual and more often than not, the primary carer is the spouse (Kessler, 1993). Although there is a wealth of literature investigating the role of dementia family care-givers (e.g. Maslach, 1981; Flicker, 1992; Murray et al, 1997), the symptoms and genetic nature of HD makes this family carer role distinct from general dementia care-giving. For example, Power (1982) notes how cognitive dysfunction in HD can lead to the patient becoming apathetic and inactive preferring to say at home, which places a huge burden on the carer. Power also found that the movement disorder associated with HD, can make families feel embarrassed to go out with the patient. Hayden et al (1980) also recognise the burden that HD places on the family and Dura (1993) notes that although educational interventions can reduce HD caregiver stress in the short term, the long term effects of intervention are minimal due to the stressfulness of continuing to provide care in this insidious and chronic disorder.

Hans & Gilmore (1968) note the major emotional, social and financial problems that caregiving in HD creates for the family, and that such issues are made worse due to lack of attention that HD has received from public health services in terms of interventions. This may be because the physical, neurological, psychiatric and genetic elements of HD mean there are no boundaries between the medical disciplines in relation to who should care for these individuals. Therefore, HD sits uncomfortably within the structure of community based services. This can have implications for the professionals involved and also HD families. Patients and their families find enormous difficulty in gaining access to specific services and professionals may not always be trained to deal with such family

dynamics. Service provision for HD families is therefore often poor and unsuitable so families are mostly burdened with the main responsibility of care (Shakespeare & Anderson, 1993).

Stress, daily hassles and psychological morbidity are often associated with family caregiving in dementia (e.g. Kinney & Stevens, 1989; Waltrowicz et al, 1996; Cousins et al, 2002). HD family carers also experience many of these problems. A number of studies have noted the psychosocial effects of HD on the family (Bolt, 1970; Davenport & Muncy, 1916; Dewhurst, Oliver et al, 1970; Hans & Gilmore, 1968; Hayden et al, 1980; Oliver & Dewhurst, 1969; Teltscher & Davies, 1972, Wallace, 1972, Yale, 1971). For example, Korer and Fitzsimmons (1985) found that the emotional and physical demands that an HD patient places upon their family can make caregiving difficult. Furthermore, lack of finances, often due to either the patient or carer (or both) having to give up their job means that there is not enough money to employ extra help to alleviate this situation. Semple (1995) carried out a qualitative study to explore and describe the experiences of family member of individuals with HD and found family carers experience a wide range of negative emotions as a result of their caregiving role and this has a significant impact on their well-being.

However, due to both the genetic implications and chronic nature of HD, family carers may experience more intense problems than dementia carers *per se* when caring for a relative with HD. Tyler et al (1983) examined the relationship between HD disease state and family breakdown and stress in a sample of ninety-two patients. Tyler and colleagues found that violence, promiscuity, bizarre and slovenly behaviour (i.e. behavioural

manifestations of HD) were often reported to be the cause of marital breakdown in HD. Behavioural problems were also cited as one of the main causes of stress within the family, with dangerous and aggressive behaviour reported in nearly half of all patients and eighty-two percent of primary carers reporting feeling stressed. Wives also reported feelings of conflict in choosing between caring for their HD affected spouse and their children over the duration of the illness. Furthermore, Hans & Koeppen (1980) argue that HD permeates the entire life of the non-HD spouse (e.g. lifestyle, family responsibility, goals and marital relationships) and so they experience continuous trauma. They found that once a diagnosis had been made, the spouse was often called upon to help in the management of the patient in terms of supervision, moral support, nursing, handling of finances and total responsibility for the home and any children.

Although such issues can be related to caregiving in many types of dementia, there are also a number of other salient factors which demonstrate that HD as a disease imposes a unique burden on family and especially spousal carers. The mood and behavioural changes associated with HD can drastically alter family, and especially spousal, relationships. Hayden et al (1980) established that in HD, the non-HD spouse has unique concerns and needs in terms of chronic isolation. They found that the anti-social behaviour associated with HD might cause social embarrassment to the carer and rejection by friends. Moreover, in a qualitative study of fifteen wives of individuals with HD, Hans & Koeppen (1980) found that partners frequently describe the way in which they feel they have ended up married to a different person and perhaps not the sort of person they would have chosen. Feelings of regret, anger and ambivalence are commonplace and often marriages come under extreme pressure. They also note that none of the partners knew of the presence of HD in the

family prior to marriage and they reacted with disbelief and denial on hearing the diagnosis. Furthermore, as the partners became aware of the steady progression of the disease process and the threat of disease transmission to any children, they became resentful and hostile. The strain on members of the family is therefore, further intensified by the impact of the unique implications stemming from the inherited nature of the disease (Williams et al, 2000). Because of the genetic implications, HD repeats itself in successive generations and once a HD patient and their spouse have had children the impact on the family may span over a number of generations if any children are found to have the disease. The availability of a predictive test to identify offspring who are at risk of developing the disease also brings its own problems in terms of the psychosocial impact it has on both the patient and their carer (e.g. Kessler, 1988; Sobel & Brookes Cowan, 2000). Often those who are 'at risk' or know they carry the gene are involved in the care of their parents or other members of their family, and are constantly reminded of the reality of HD. It is not uncommon for a person to nurse their parent, then an older sibling and finally succumb to HD themselves, whilst worrying all the time that they have transmitted the disease to their children (Kessler, 1993).

Despite these issues "little or no professional attention has been given to [carers] in the HD literature" (Kessler, 1993: 145). Furthermore, the majority of studies in existence are relatively small scale and qualitative in nature, making it hard to generalize findings beyond the sample population itself. As there is currently no cure, it is not surprising that it is the patient and those who are 'at risk', who receive the most attention with only a few prominent papers discussing the impact of HD on the family carer (e.g. Hans & Koeppen, 1980; Kessler, 1993; Tyler et al 1983). However, this does leave a clear gap in the literature in which to further investigate the impact of HD on the QoL of family carers.

Motivation for Research.

Generally, the psychologist's role in HD has traditionally been one of assessment, management, evaluation and research into the disease processes and the sufferer themselves rather than the carer. Numerous scales are used to assess the severity of symptoms (e.g. Stroop, 1935, and Wisconsin (Nelson, 1976)) which can demonstrate very early changes in attention, ability to learn etc, enabling diagnosis often prior to the onset of chorea. These are clearly useful in preparing control and experimental groups in research but, are of less practical value to the sufferer themselves and their family. Therefore HD affected families may experience a lack of expertise and specialism from health professionals on practical aspects such as therapeutic interventions, advice on genetic counselling or continuity of care.

As with many diseases where there is no cure, focus is quite rightly and obviously placed upon finding a successful treatment. Since the discovery of the Huntintin gene (Gusella et al, 1983) patient care has changed quite dramatically in terms of both searching for a cure and developing more appropriate and specialised care facilities. However, carer issues still appear to remain constant with current literature highlighting problems that were raised in the 1920's. It is therefore clearly important to put resources into establishing methods of alleviating the carer burden in HD by successfully addressing carers' needs.

With the general remit of Health Psychology being to promote and maintain wellbeing via the application of psychological models and theories, the issue of caregiving in HD is undoubtedly an area in which the Health Psychologist can have an impact. For example,

through the practical application of theory such as designing self-care programs, using therapeutic interventions or by advocating carers' views on service quality. However, with health care purchasers increasingly being expected to allocate resources on the basis of the evidence of the effectiveness of health care interventions, lack of systematic research poses a huge problem. Therefore, one of the most pressing issues is the generation of quality research with which to test theories and interventions for HD family caregivers.

Because of the genetic implications of HD, it impacts upon individual family members in different ways. Spousal carers who have children have to deal with the possibility that their children may also be carrying the HD gene. This puts them in a position where they could be caring for affected loved ones over a number of generations on top of having to cope with the possibility of resentment towards their spouse for putting them in this situation. Children and other family members who are carers may have to cope with either an 'at-risk', HD gene-positive or HD-gene negative status thus giving them the possibility of watching how they themselves may deteriorate in the future or what they have been saved from. These different carer roles are therefore very distinct with regards to the burden that they may place onto individual carers. Therefore, in order to develop a well validated tool, it was decided that the current studies would focus solely upon spousal carers with the aim of revalidating the scale at a later date for different sub-populations of HD carers.

This current research therefore, aimed to establish the factors that enhance and compromise the lives of HD spousal caregivers by utilising the theoretical construct of quality of life (QoL). Since the 1960's, QoL has been emerging as a useful outcome measure by which to

judge the efficacy of psychological interventions (Rapley, 2003). It was hoped that by using QoL as a measure of outcome for HD caregivers, attention would be focussed upon the impact of HD, the individual carer's emotional and physical functioning and lifestyle. Therefore, such QoL indicators may ultimately help to answer the question of whether an intervention leads to an increase in wellbeing by providing a carer-led baseline against which the effects of the intervention can be evaluated. With this in mind, a disease-specific measure was developed to bring together theoretical constructs and practical application and produce a user-friendly QoL measurement for HD spousal carers that could be used to implement and assess therapeutic interventions.

Research Aims.

1. To examine the construct of QoL with HD spousal carers and Health Care Professionals working in the field in order to investigate whether the construct itself is meaningful and relevant to the HD spousal carer.

2. To obtain a detailed understanding of the issues surrounding QoL and spousal caregiving in HD, through the use of photographs and carer's own comments.

3. To take findings from studies 1 and 2 to focus groups for discussion in order to clarify participants' comprehension of terminology and concepts developed during the preliminary investigations.

4. To develop a disease-specific QoL measure for spousal carers of HD patients (HDQoL-C).

5. To validate the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) through pilot investigation.

Chapter 5.

Exploratory Study 1: Is the concept of quality of life important to Spousal carers of Huntington's Disease patients?

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Background.

There is very little literature on spousal caregiving in HD and no specific literature that focuses on the specific concept of QoL. It was therefore felt necessary to initially establish that QoL was indeed a concept that was pertinent to spousal carers. In addition, it was important to establish whether health care professionals felt that HD spousal carers' QoL was being compromised in any way in order to try and 1) get an idea of the 'bigger picture' and 2) investigate whether there was concordance between the views and perceptions of carers and health care professionals. Therefore, in this initial study the domains and facets of the Comprehensive Quality of Life Scale for Adults (ComQol-A5; Cummins, 1997) were rated by using a likert type scale in order to achieve two main objectives: (1) to examine the relevance of the ComQol-A5 domains and facets to the perception of HD Spousal Carers and (2) to consider the development of any additional facets and remove less relevant facets. Carers and Health care professionals were also asked to write down any issues that they felt were pertinent to their QoL as primary carers of HD patients. Analysis of the ratings data revealed that 25% or more of answers in each facet fell into response brackets 4 and 5 ('very important and extremely important') This means that at least 25% of all participants perceived every facet as either very important or extremely important to their quality of life. Further analysis of the qualitative data established 18 sub-themes relating to quality of life which clustered into four final themes of Professional Issues in HD, Personal Wellbeing in HD, Practical Issues in HD and Emotional Wellbeing in HD.

Method.

Sample:

Family carers and health care professionals were recruited though a research talk at the Huntington's Disease Association's Annual General Meeting (Summer, 2001) and asked to take part in an investigation into quality of life issues in Huntington's Disease (HD). Twenty HD spousal carers (6 males and 14 females; mean age, 52.3 years, sd = 2.62) and twenty health care professionals (4 males and 16 females; mean age 49.4 years, sd = 3.45) agreed to take part in this initial study. Carers spent between 12 and 17 hours per day caring for their affected spouse and as such, caregiving was deemed to be their full-time occupation. Carers' educational background ranged from secondary school to post-graduate level, previous employment details were not recorded. Health care professionals were a mix of care advisers (n=4), nurses (n=10), genetics specialists (n=2), social workers (n=2) and senior management (n=2). They were all educated to at least undergraduate university level.

Materials:

The underlying concepts and procedures for assessing QoL were based on Cummins (1997) Comprehensive Quality of Life scale - Adult (ComQol-A5). This is a comprehensive, multidimensional and well validated quality of life measurement, consisting of 35 facets subsumed in 7 domains of Material Wellbeing, Health, Productivity, Intimacy, Safety, Place in the Community and Emotional wellbeing, designed for use with the general adult population (See appendix IV). Using a 7-point Likert scale, participants are required to tick

the box that most closely reflects how they feel about various areas of their life (1= delighted, 7 = terrible).

Procedure:

Spousal carers and health care professionals were recruited through a research talk at the Huntington's Disease Assocation (AGM) (Summer, 2001). QoL was operationalised to the participants using the following definition:

"Quality of life is both objective and subjective, each axis being the aggregate of several domains: material well being, health, productivity, intimacy, safety, community and emotional well being. Objective domains comprise culturally-relevant measures of objective well being. Subjective domains comprise domain satisfaction weighted by their importance to the individual." (Cummins, 1997, pg 7).

Participants did not answer the QoL scale but instead rated each item for its relevance to a HD spousal carer. Answer sheets with 5-point likert response scales were explained and participants were asked to tick the most relevant category (see appendix V).

In addition, in order to gain qualitative data on the participants' evaluation of the ComQoL-A5 and to provide an opportunity to describe any other issues pertinent to HD, participants were also asked to write down what they felt were the main issues surrounding the QoL of primary carers of HD patients and make further comments in relation to the questionnaire itself. All participants gave informed written consent and were advised of their right to withdraw from the study if they wished to do so.

Findings.

Ratings Data:

Due to the exploratory nature of the study, the ratings data were analysed purely at a descriptive level, using percentages. The structure of this preliminary investigation was adapted from Jirojanakul & Skevington (2000).

The data obtained from the carers and health care professionals in relation to the relevance of each facet (or aspect of life) on the COMQ-L to the QoL of HD spousal carers revealed that 25% or more of answers in each facet fell into response brackets 4 and 5 ('very important and extremely important') This means that at least 25% of all participants perceived every facet as either very important or extremely important to their quality of life.

Analysis of overall domain scores revealed that some facets on the Objective domains of 'Material well being', 'Health', 'Productivity' and 'Place in the community' were considered to be of little relevance to HD spousal carers' QoL. Subsequent item analysis established that at least 50% of responses to the following items fell into the response scales 1 and 2 ('not at all important' and 'not very important'), see Table 5.1 below.

Table 5.1. Objective Facet Items considered 'not at all important' or 'not very important' to HD spousal carers QoL.

Q1 b) How many personal possessions do you have compared to other people?
Q2 a) How many times have you visited the doctor in the last 3 months?
Q2 c) What regular medication do you take each day?
Q3 c) On average, how many hours of TV do you watch a day?
Q6 b) Do you hold an unpaid position of responsibility in relation to any club, group, Or Society?
Q6 c) How often do people outside your home ask for help and advice?

Within the Domains of 'Importance' and 'Satisfaction' all items with the exception of Q2 (1) *'How important to you are the things you own'* and Q 3 (1) *'How satisfied are you with the things you own'* were considered to be relevant to HD spousal carers' QoL. Subsequent item analysis established that at least 50% of responses to the following items fell into the response scales 4 and 5 ('very important' and 'extremely important', see Table 5.2. below).

Table 5.2. Subjective Facet Items considered 'very important' or 'extremely important' to HD spousal carers' QoL.

Q 2. HOW IMPORTANT TO YOU...	... .is your Health? ... .is what you achieve in life? ... .are close relationships with family and friends? ... .is how safe you feel? ... .is doing things with people outside your home ... .is your own happiness?
Q3. HOW SATISFIED ARE YOU...	... .with your health? ... .with what you achieve in life? ... .with your close relationships? ... .with how safe you feel? ... .with doing things with people outside your home? ... .with your own happiness?

Further analysis of question 2.1 (*'How important to you are the things you own')* and question 3.1 (*'How satisfied are you with the things you own')* established that at least 50% of responses to the following items fell into the response scales 1 and 2 ('not at all important' and 'not very important').

Correspondence between the views of carers and health care professionals.

Both carers and health care professionals had corresponding opinions about the relevance of each domain and facet. For both groups, highly relevant QoL items included questions such as, *how often do you sleep well?; How often do you do the things you really want to? If you are feeling sad or depressed, how often does someone show they care for you?* Less relevant items included questions such

as, *How many personal possessions do you have compared with other people?; On average, how many hours of TV do you watch each day?; How satisfied are you with the things you own?*

Interpretation of Ratings Data:

Importance ratings for objective and subjective domains were consistent across the domains of 'Material well being', 'Intimacy', 'Safety' and 'Emotional well being' but inconsistent in relation to the domains of 'Health', 'Productivity' and 'place in the community'. As such, facets from these three domains and the additional facets that were considered of little importance to QoL were noted for subsequent discussion within a focus group setting in order to establish their inclusion / exclusion in the pilot questionnaire.

Qualitative Data:

Fifteen carers highlighted a number of issues that they felt were the main concerns surrounding the QoL of primary carers of HD patients.

The 256 comments noted were investigated using a phenomenological approach to establish the 'perceived meaning' within the text rather than 'objective reality'. Interpretative Phenomenological Analysis (IPA) (Smith, 1995; Smith & Osbourn, 2004) was adapted for use with this data. The data consisted of a combination of paragraphs, sentences and words from a number of carers. Ordinarily with IPA, each transcript would be taken person by person in order to capture the essence of each person's perspective. However, due to the

exploratory nature of the study and the small amount of data involved, the comments were analysed as if one continuous script.

Analysis:

The meaning of the comments was central to the analysis. As such the transcript was read a number of times in order to get a feel for the data and become familiar with the comments made. Issues that the participants raised were annotated in the text and similarities / differences between comments were noted. Once this had been completed for the whole transcript, initial annotations were transformed into direct phrases with the aim of capturing the meaning of what was found in the text. The emerging sub-themes were then investigated for connections between them and as such, were clustered together into the final themes. As the final themes emerged, they were related back to the transcript in order to ensure that the connections worked for the primary source material.

Using this method 18 sub-themes emerged directly from the raw data which clustered into the following four final themes of 'Levels of support', 'Dissatisfaction with caregiving role', 'Practical aspects of caregiving' and 'Feelings and emotional wellbeing'. These themes were agreed by two independent raters (intercoder reliability K = 0.8). See Table 5.3 below.

Table 5.3. Themes and sub-themes generated from concept clarification data.

THEMES	SUB-THEMES
Levels of support	Appropriate help from social services Professional knowledge and understanding Appropriate specialist services Appropriate care facilities Support from health care professionals
Dissatisfaction with caregiving role	Duty of care Benefit of personal support Advocacy Burden of Responsibility Genetic Issues Loss of Identity
Practical aspects of caregiving	Safety and Security Practical support Financial Burden Tiredness Lack of time Support from friends and family
Feelings and emotional wellbeing	Loss of emotional closeness Isolation Negative emotions Future concerns

Participants also commented on the design and format of Cummins' (1997) questionnaire. Utilising the same adaptation of IPA as with the previous data, three separate issues were identified from the 54 comments made. The main issues raised by participants were that it was important to devise a scale that was short, easy to understand and used a likert scale format.

Interpretation of Qualitative Data:

These exploratory findings suggest that that there is an interrelationship between the facets on the ComQoL-A5 (Cummins, 1997) and the final themes and sub-themes that emerged from this initial data. The sub-themes of *personal support*, *safety and security*, *practical support*, *financial burden*, *time* and *support from friends and family* are all identifiable facets already included in the ComQoL-A5. However, a number of disease-specific issues in relation to both caregiving and HD itself were evident in the comments noted by the participants in this study and these themes are discussed individually below.

Levels of support:

The theme *'Levels of support'* especially highlighted areas that are mainly applicable to people who either have or are caring for someone with a chronic illness. The sub-themes of *'appropriate help from social services'*, *'professional knowledge and understanding'*, *'appropriate specialist service'*, *'appropriate care facilities'*, and *'support from health care professionals'* are not issues that would normally concern the general population. These issues also appear to be exacerbated due to the chronic nature of HD and the ongoing frustration of not being able to get the specialised help required as a HD caregiver.

Dissatisfaction with caregiving role:

Within the theme of *'Satisfaction with life'* the sub-themes *"duty of care'*, *'benefit of personal support'*, *'burden of responsibility'* and *'loss of identity'* link well with the current caregiving literature (e.g. Kessler, 1993). However, particularly notable were the sub-themes such as *'advocacy'* and *'genetic issues'* in which carers commented upon the difficulties of advocating for someone

with HD and made note of how devastating the impact of the genetic implications of HD are upon the carer and the whole family.

Practical aspects of caregiving:

The theme of *'Practical aspects of caregiving'* encompassed a number of issues that are relevant to the general population as already mentioned above. However, sub-themes such as *'practical support'* and *'lack of time'* are once again indicative of a chronic caregiving experience.

Feelings and Emotional Wellbeing:

The final theme of *' Feelings and Emotional Wellbeing'* highlighted the chronic nature of caregiving in HD with sub-themes of *'loss of emotional closeness'*, *'Isolation'*, *'negative emotions' and 'future concerns'* all relating directly to the specific nature of the HD in terms of both the nature of the disease, the affect that it has on the sufferer's personality and the progression of the illness.

Conclusions:

This study provides preliminary evidence that spousal carers of HD patients and health care professionals would value a disease-specific quality of life scale that could be used to evaluate their objective and subjective QoL. The generic quality of life scale (Cummins, 1997) was relevant to the QoL of spousal carers, although it was apparent that many of the issues and concerns that HD spousal carers have, were not being 'tapped in' to though the generic

questionnaire. Furthermore, there were a number of elements in the generic questionnaire that were felt unimportant or irrelevant to the QoL of HD spousal carers.

As the qualitative data from this study was both minimal and tentative, it was appropriate to carry out a more in-depth analysis of the impact of HD on the QoL of spousal carers. This was carried out using 'photovoice' as a method in which spousal carers could take photographs and make comments on aspects of things in their lives that are important to them.

Chapter 6.

Exploratory Study 2: Capturing the Huntington's Disease spousal carer experience using the 'Photovoice' method.

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Background.

Study 1 provides initial evidence that QoL is an important concept to HD spousal carers. However, previous research into the experience of spousal caregiving in HD is too sparse to make too many assumptions. Furthermore the concept of QoL is difficult to operationalise (Cummins, 1997; Rapley, 2003). The objective of this second exploratory study therefore, was to capture and describe the experiences of HD spousal carers specifically in relation to their QoL using an in-depth and insightful methodology. In order to gain insight into the complex role of the carer, visual representations of QoL and corresponding written information were gathered using 'Photovoice'. This is usually a process by which people can identify, represent and enhance a specific community through photography (Wang, 1999). However, for the purpose of this study 'Photovoice' was employed as an opportunity for individual participants to explore the concept of QoL by photographing and giving written reflections on specific QoL issues surrounding their care giving and HD. Previous health psychology research has noted the benefits of using photography in order to produce rich and informative data (Radley, 2001). With this in mind it was hoped that by combining the objective image created by the photograph with the subjective account of the meaning behind the picture, the meaning of the image would be 'anchored down' by the participant rather than the researcher imposing meaning onto the photographs. Five spousal carers photographed and described elements of their life in which they felt their QoL was being enhanced or compromised and the data was analysed using basic content analysis (Weber, 1990). Using content analysis, nine manifest themes were identified and tentative latent inferences were made in relation to these themes. Although some positive issues did

emerge, these were minimal compared to the negative impact that HD had on carers' overall QoL. Seven out of the nine themes that emerged were also evident in at least one of the seven QoL domains on the Comprehensive Quality of Life scale - Adult Version (ComQoL-A5; Cummins, 1997); suggesting that QoL is negatively affected for these spousal carers.

Method.

Sample:

Participants were recruited through the Huntington's Disease Association UK. Five family carers, four females and one male (mean age 49.1, SD = 2.54 years) volunteered to take part. No other sample characteristics for these participants were sought; however, the sample were predominantly white and British. All participants gave informed written consent for their photographs and corresponding written data to be released for publication and were aware of their right to withdraw from the study at any time.

Materials and Procedure:

Carers were provided with disposable 27-exposure, colour film cameras and a corresponding dialogue sheet. They were given a written description of Cummins (1997) and the World Health Organisation's (WHO, 1995) definitions of QoL i.e.

"Quality of life is both objective and subjective, each axis being the aggregate of several domains: material well being, health, productivity, intimacy, safety, community and emotional well being. Objective domains comprise

culturally-relevant measures of objective well being. Subjective domains comprise domain satisfaction weighted by their importance to the individual." (Cummins, 1997, pg 7).

"... .an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns". (WHO Group, 1995, pg 3).

Cummins' (1997) definition was utilized because it attempts to define QoL as a global construct. It provides a positive perspective to life quality and psychological research as it encompasses both positive and negative dimensions of QoL. It further accounts for both objective and subjective interpretations of QoL, which is a very important consideration when explaining individual experiences in terms of well-being.

Carers were subsequently asked to use their disposable cameras to take photographs of things (not people) that represented a compromise or enhancement of their QoL. They were told that they did not have to use all of the film in the camera and that it did not matter if they took more pictures of enhancing things rather than compromising things, or vice versa. They were told that what mattered was the pictures they took were significant to them. Furthermore, straight after they had taken a photo they were asked to write a sentence or two about it. All photos were taken within a two-week period and the cameras returned for development. Negatives were returned to participants so they could identify the pictures that were most important to them. It was hoped that this would engage carers in a process of reflection by addressing what the photographs conveyed and what this meant to them.

Analysis:

Using basic content analysis (Weber, 1990) 109 photographs were analysed for manifest content and classified into nine themes representing differing aspects of the carer experience in relation to QoL. For each photograph, the participant wrote a sentence or two about why they had taken the picture and what it meant to them. This corresponding text allowed for tentative latent inferences (as described by Tashakkoir & Teddlie, 1998) to be made with regards to each photo, and ultimately each theme. The themes were then compared to the identified themes within the domains of the ComQoL-A5 (Cummins, 1997).

Findings.

109 photographs and their corresponding text were classified into nine themes representing differing aspects of the carer experience in relation to QoL. The photographs, corresponding text and theme classifications were re-analysed by a second researcher (intercoder reliability $K = 0.8$). Table 6.1 shows the nine themes identified, the number of photographs classified into each theme and an example from the dialogue sheet.

Table 6.1. Identified themes and example quotes from the 'photovoice' data.

Theme	Number of Photos	Quote
Care and security	16	"We are cared for by our church"
Small pleasures	11	"A cigarette is one of the main pleasures in life"
Loneliness	13	"No-one to talk to"
Escape	8	"There is no space just to be"
Sense of loss	11	"Families lose so many members to the same disease"
Neglected Needs	10	"We miss out on all sorts of things"
Support	17	"The logo of our charity enables us to recognise other people who suffer with the same affliction"
Time	12	"Dash home from work, the Solpadol and coffee routine, off to the supermarket. We rarely eat before 9pm".
Daily Hassles	11	"We have to wash his bedding 3 - 4 times a week, I long for a day without washing!"

The themes identified are discussed individually below, including excerpts from the dialogue sheets and photographs taken. It can be tentatively suggested from these excerpts that the majority of the dialogue within the data is associated with negative factors which impact on the carers' lives. They describe a life which is dominated by their care-giving role, to the detriment of their own needs.

Care and Security

Carers describe situations in which small things can ease their mind and make them feel more secure.

Image 1.

Comment: *"Making the house the way we want it now will help in the future. It will also be a safe place for our family, a place that my children can always come home, a place where they are loved and safe"*

Small Pleasures

Carers take great satisfaction in the small things in life in order to enhance their day to day living.

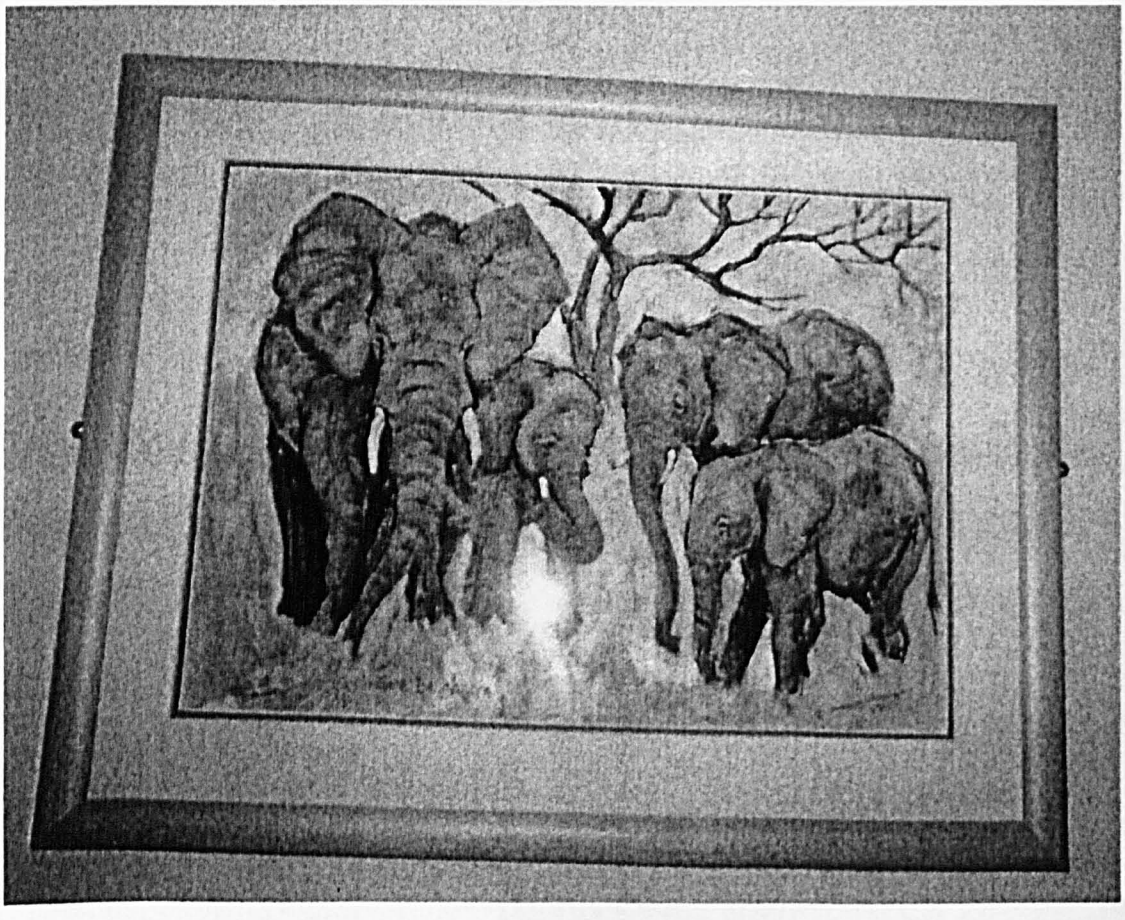
Image 2.

Comment: *"Flowers in the house are my bit of luxury, all of my friends know this. These are a present from X (HD affected individual). The significance of the delicate beauty and frailness; the short lifespan but the pleasure they bring – Huntington's Disease"*

Loneliness

Isolation is a significant problem for carers, with many of them not having the time to socialise with their friends or even stay on the phone for too long.

Image 3.



Comment: *"I have a picture of the year we met. The colours in the background, the pinks, contain the colours I see when meditating. A sense of loneliness perhaps"*

Escape

The need to escape from their situation combined with the reality that there is no release is evident in the carers' dialogue.

Image 4.

Comment: *"One half of the rainbow from our terrace gives a feeling of space and light at the end of the tunnel. Will she ever die? Will we be able to have some life? Quality of life is elusive, like the pot of gold at the end of the rainbow"*

Sense of Loss

Carers recognise a sense of loss. Not only the loss of a spouse, but also the potential loss of a fit and healthy family.

Image 5.

Comment: *"Toys in the garden. With a genetic disease it means that we choose not to have children – our future is taken away from us"*

Neglected Needs

Carers describe the constant interruptions they encounter from living with HD. They want the ability to fulfil their own needs and wish for a simpler life.

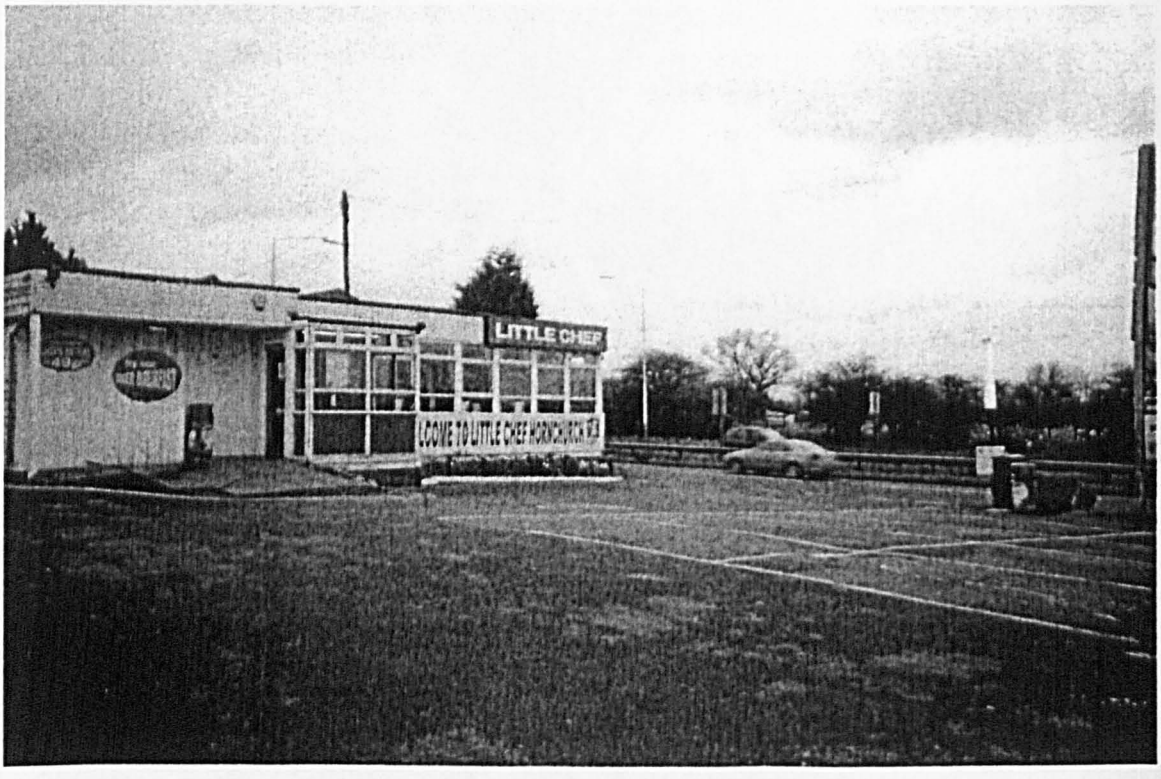
Image 6

Comment: *"Every time I go shopping with X (HD affected individual) it's so tedious trying to remember what I want. In fact it's impossible. I try and get everything she needs just in case she runs out- what about my shopping? What about my needs?"*

Support

Carers recognise and appreciate the small amount of support that they receive from different sources.

Image 7.



Comment: *"Most people are helpful when in contact with a disability. The waitress at Y restaurant was brilliant at dealing with HD impatience – although not knowing what was wrong"*

However, they also noted a lack of support from a number of service providers.

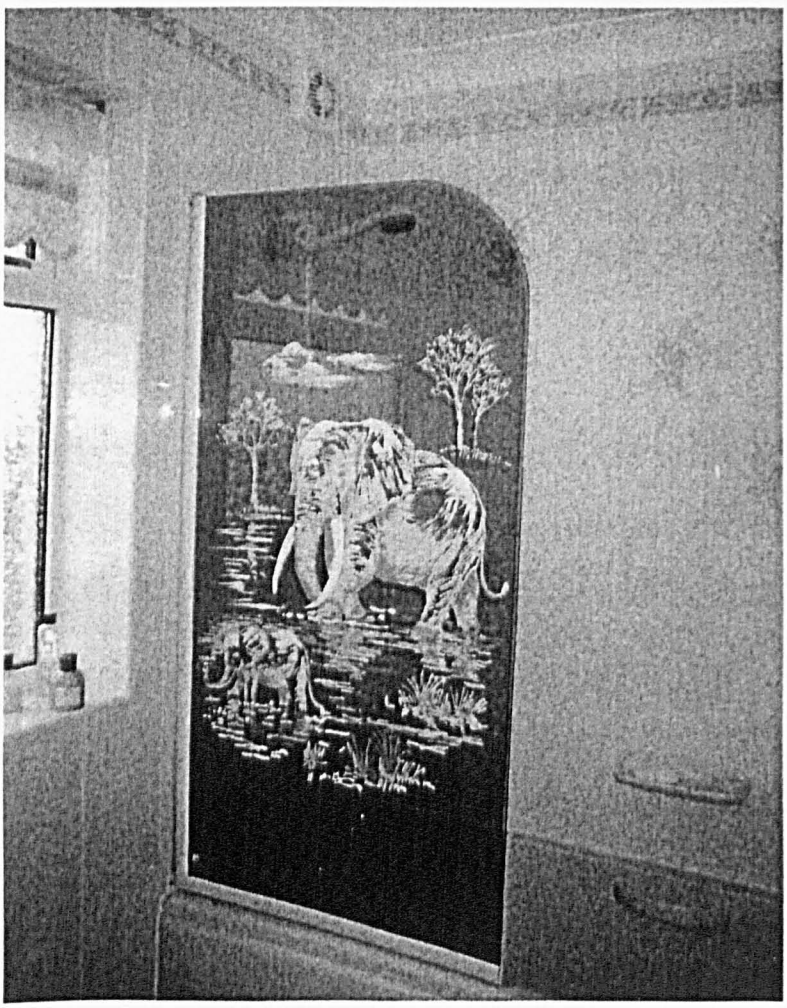
Image 8.

"Requests for a psychiatric assessment were not acted upon and we were left in a high stress situation".

Lack of time

Lack of time is a big issue for HD carers. They find themselves continually compromised between doing things for their affected relative, and finding time for themselves.

Image 9.

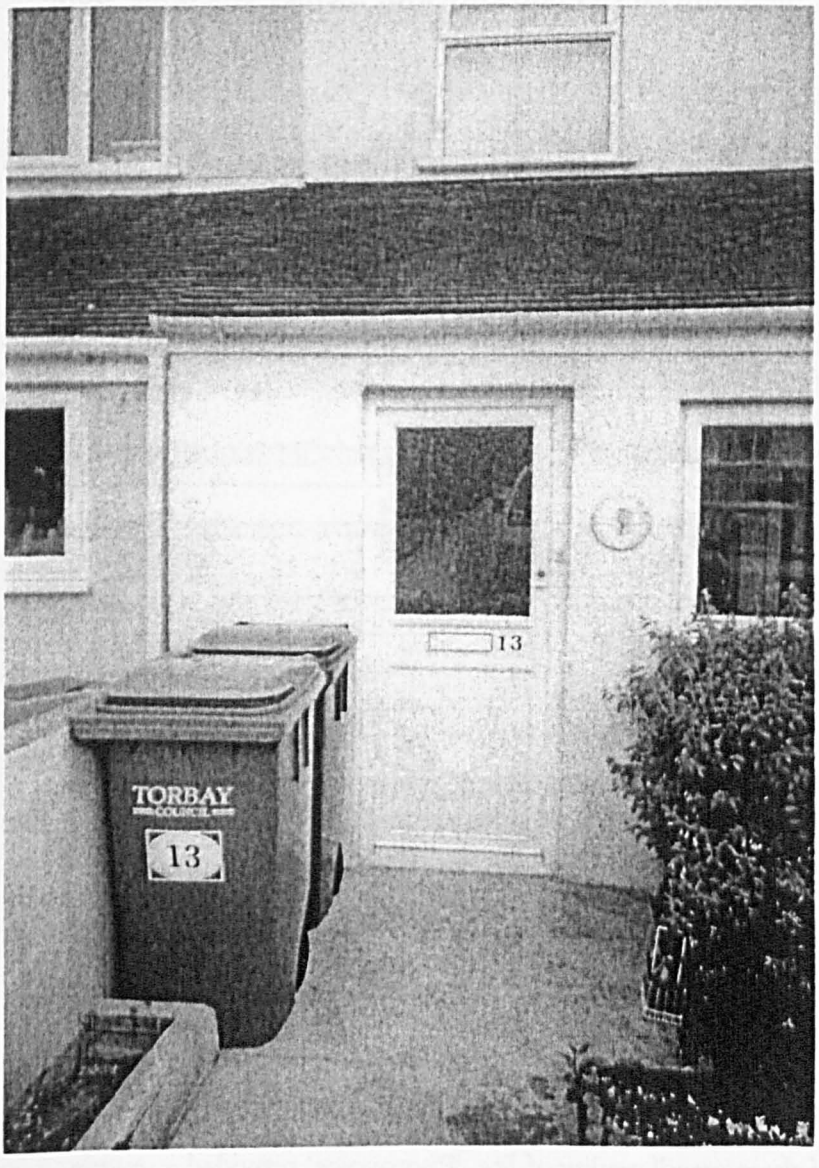


Comment: *"I bought a shower screen over 15 years ago. I had stored it and when we had saved enough to do the bathroom we spent over 2 years trying to replace the perished fixings. With HD it's always a question of time, you can't follow things through"*

Daily Hassles

Carers further describe the way in which the hassles of daily living impact upon their lives.

Image 10.



Comment: *"The Wheelie Bins. We need extra because of all the stuff she throws away. We could feed another household I'm sure. She always misjudges and you can't reason with her. I could weep, it's a vicious circle"*

These exploratory findings suggest that that there is some overlap between the domains on the ComQoL-A5 (Cummins, 1997) and the final themes that emerged from this photovoice data. Table 6.2 below compares the domains of the ComQol-A5 to the themes that were identified in this study.

Table 6.2 Overlap between the ComQoL-A5 domains with themes emerging from the photovoice data.

COMQoL-A5 Domains	Themes from Photovoice data
Material Well-being	*
Productivity	Lack of Time
Intimacy	Care and Security Sense of Loss Support Loneliness
Safety	Care and Security
Place in the community	Neglected Needs
Emotional Well-being	Neglected Needs Escape
Health	*
*	Small Pleasures
*	Daily Hassles

* No overlap between the ComQol-A5 domains and the Photovoice themes.

Interpretation of Findings.

The aim of this preliminary study was to explore and describe the experience of HD spousal care-givers in terms of their overall quality of life. 'Photovoice' provided a unique opportunity for the HD carers to capture and reflect on issues as they arose (and became apparent as influencing their QoL) on a day-to-day basis. This may not have been

possible using other investigative methods. In addition, Photovoice' gave carers the opportunity to reflect on their carer role, but required only minimal time out from their caring routines. This is an important issue, as lack of time was identified as a theme in this data and is acknowledged in the care-giving literature (Waltrowicz et al, 1996).

Analysis of the photovoice data produced nine themes that appear to be intrinsically linked with HD spousal carers' QoL. The identified themes lean towards the notion that caring for a family member with HD imposes a unique and difficult burden on the carer's life. Although some positive elements emerged, these appeared to be minimal (i.e. small pleasures like having a cigarette or having flowers in the house) when compared to the negative impact that HD has on their lives. We found that these informal carers often experienced loneliness, a need to escape and a unique sense of loss while trying to adequately care for their loved ones and maintain some form of QoL for themselves. Such issues are unique for HD spousal carers as due to the genetic nature of the disease, there can often be no end to their caregiving role unless they make a conscious choice not to have children. Previous research in relation to each of the themes is discussed more explicitly below.

The themes 'Care and security' and 'Small pleasures' that emerged in this data are not readily identifiable within in the existing HD caregiving literature. Within the theme 'Care and Security' carers described how small things such as making the house safe and secure were a real comfort to them. Furthermore, within the theme of 'Small Pleasures', carers commented upon how the little things in life such as having an empty washing

basket or fresh flowers in the house became very important and precious to them. It could therefore, be tentatively suggested that using a method such as 'Photovoice' has allowed us to tap in to very explicit and individual areas of concern for these carers as they considered the issues that affect their QoL.

The theme 'Loneliness' was referred to frequently amongst the carers in this study. They reported feeling isolated from their friends and other social networks. Murray (1995) and Murray et al (1997) have also noted that spousal carers of dementia patients experience isolation and further, that they feel obligated to spend much of their time at home. Furthermore, Hayden et al (1980) note how the anti-social behaviour often associated with HD can lead to a decision on behalf of the HD spouse to isolate him/herself in order to avoid embarrassing situations.

The theme 'Escape' encompassed a number of issues that highlight the HD spousal carers situation. Carers commented that they felt there was no way out and no end to their caregiving role, and wrote about a chronic and difficult burden that they felt was imposed onto them. Flicker (1992) also established that caring for a family member with a dementing illness imposes an intolerable burden on the family as they carry out their caregiving role. Furthermore Roos et al, (1990) argue that facing a 'frightening future' is one of the causes of difficulties for partners of HD patients.

The theme of 'Sense of loss' that emerged within this study was primarily linked to the genetic nature of HD for these carers. The unique nature of caring for someone with HD was illustrated by carers expressing their loss as losing the opportunity to have children

and early deaths generally within the family; not solely the inevitability of losing their spouse. Previous research has shown that the strain on family members appears to be intensified by the impact of the implications stemming from the inherited nature of HD and further, by the availability of a predictive test to identify offspring who are at risk of developing the disease (Kessler, 1987; Huggins et al., 1992).

The theme of 'Neglected needs' that emerged within this study, incorporated evidence of carers talking explicitly about how their own needs are compromised in order to care for their spouse. This reflects Kessler's (1993) argument that spouses often neglect their own needs in order to shoulder the burden of decision making, nurturing, caring and wage earning. Maslach (1981) and Tyler et al (1983) also describe the hardship that is often experienced by informal carergivers. Such hardship was also evidenced by the carers in this study who reported both financial and emotional hardship in terms of their neglected needs.

The theme of 'Support' that emerged from this data is also evident in a number of previous studies. For example, Waltrowicz et al (1996) note that service providers who deal with dementia sufferers and their families appear to have an incomplete impression of the experience of the carer, as informal carers complain infrequently to professionals about their own problems. Furthermore, Tyler et al (1983) note that family support is an essential factor in any long-term alleviation of the stressful situation that HD families find themselves in. Dura (1993) further argue that HD carers require continuous support in order to reduce the burden that caregiving brings. Shakespeare and Anderson (1993) found that members of HD families often criticise the lack of services available to them.

Interestingly, in this study we also found that lack of support from service providers was reported by the carers. However, the carers in this study placed a positive emphasis on the support that they did receive from the family and community and noted how important this was to them in facilitating their caregiving role.

The theme of 'Time' was a big issue for carers who found themselves torn between caring for their spouse, looking after their children and finding some time for themselves. Waltrowicz et al (1996) note that carers who are children of dementia sufferers are often torn between caring for an elderly parent and the welfare of their own children and spouse. Kessler (1993) notes that spouses of HD patients with young children tend to find conflict over their loyalty towards their partner and their obligation to their children. Furthermore, Hans and Koeppen (1980) note how HD permeates the entire life of the spousal carer.

The theme of 'Daily Hassles' was evident within this data. However, this is not addressed in its entirety within the existing HD caregiving literature. Inferences in relation to coping strategies and stress are numerous (e.g., Hans & Koeppen, 1980; Maslach, 1981; Tyler et al, 1983) and the impact of daily hassles for family carers of dementia patients is described by Kinney & Stevens (1989). However, the sheer impact that small tasks such as doing the shopping or washing has not been clearly highlighted in previous HD research.

Further analysis of the 'photovoice' data established that seven out of the nine themes that emerged could be related back to at least one of the seven QoL domains on the

ComQoL-A5. This suggests that QoL is indeed impacted upon in some way for these spousal carers and there is some form of psychological burden associated with the spousal caregiving role in HD. However, the themes of 'Small Pleasures' and 'Daily Hassles' were unable to be satisfactorily related back to the ComQoL-A5. Interestingly, such themes are also difficult to identify within the HD caregiving literature. It may be that using 'Photovoice' to gather rich data has helped to identify such small issues and how they can become amplified and impact greatly upon QoL in a chronic caregiving situation. However, such inferences should be taken with caution due to the small sample size and exploratory nature of this study.

None of the emerging themes from the 'Photovoice' data could be satisfactorily related to the domains of 'Material well-being' or 'Health' on the ComQoL-A5 (Cummins, 1997). In terms of 'Material well-being', findings from study 1 established that HD carers considered material possessions from the perspective of being able to provide suitable care for their affected loved one rather than concerning themselves with material possessions *per se*.. In terms of the 'Health' domain, this is interesting as previous analysis of overall domains scores on the ComQoL-A5 and HD spousal caregiving (Aubeeluck & Buchanan, 2003) established that the objective domain of 'Health' on the ComQoL-A5 was considered to be of little relevance to HD spousal carers. However, in the same investigation, subjective questions relating to the "importance of health" on the ComQoL-A5 were deemed to be very important to the carers. Certainly, existing global quality of life scales (e.g. WHOQOL, 1998, Cummins 1997) attempt to measure aspects of quality of life that are regarded as pertinent to health status, such as life satisfaction, well-being, functional ability and stress (Bowling, 1997). However, there is still debate with regards to whether this should be

measured objectively or subjectively. Researchers are increasingly leaning towards subjective self-ratings of health (Rapley, 2003). However, policy makers often prefer more objective and classifiable indicators that may ultimately be more useful in the formation of health policies. It is possible that spousal carers of HD patients only consider their health to be important in terms of their caregiving role, i.e. they are only concerned for their health when it impacts upon their ability to care for their spouse. This discrepancy between the objective domains of QoL and subjective importance of health is an issue that needs to be addressed further to try and establish the real meaning of 'Health' for spousal carers of HD patients.

It is important to recognise that this study does have limitations. Firstly, there is only a small number of carers included in the study. Secondly, although it is widely recognised in Europe that wives are the largest group of caregivers (McMurphy et al, 1993) this study does have a bias towards female spousal carers making generalisability more difficult. However, we stress that these findings are preliminary and should be viewed as such. In addition, the strengths of this study can be found in the novel methodology employed and the rich data obtained from it.

Conclusions:

This data suggests that carers struggle to maintain their sense of self. The carers in this study often neglected their own needs as their caregiving role and the disease process took over their lives as well as the life of their HD affected spouse. The results show there are some similarities with care-givers of those with other types of dementia (e.g Dura et al, 1990) but that there is also a need to consider HD independently due to the

unique nature of the disease. The change of marital role that inevitably comes with caring for a spouse with dementia is often compounded in HD by extreme isolation that frequently follows social embarrassments due to behavioural problems associated with HD. Furthermore, the non-HD spouse has to take on board the fact that HD may also have been transmitted to any children and as such they may be placed in a position of caregiving for a number of decades (Hayden et al, 1980), or may decide not to have a family.

With findings from study 1 complementing the findings from study 2, it was decided to further investigate the concept of QoL with HD spousal carers within a focus group setting. The aim of the focus groups was to investigate these emerging HD specific QoL themes with regards to spousal caregiving and further, to generate items for use in the piloting of a HD disease-specific quality of life scale.

Chapter 7.

Exploratory Study 3. Investigating emerging themes in a Focus Group setting.

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Background.

The findings from studies 1 and 2 provided evidence that QoL is both a concept that is relevant to HD spousal carers and further that their QoL is greatly impacted upon due to their caregiving role. The purpose of a third exploratory study was therefore to further investigate these findings within a larger sample of spousal caregivers and to provide a clear framework for designing a HD specific QoL measure for spousal carers. Therefore, in this third study the conceptual frame work of the initial draft of the Huntington's Disease Quality of Life Battery for Spousal Carers (HDQoL-C) was tested through the use of 6 semi-directed focus groups. The focus group is a recognised setting for gathering QoL information (WHO, 1994) and as such, the two main objectives were: (1) to further clarify disease-specific aspects of QoL that were deemed important to potential users of the questionnaire and (2) to further examine the relevance of the general QoL domains and facets for HD spousal carers. Analysis of the focus group data supported the identification of the four themes of Levels of Support; Dissatisfaction with Caregiving Role; Practical Aspects of caregiving; Feelings and Emotional Wellbeing in Study 1. All the previously identified sub-themes from studies 1 and 2 were also evident in the focus group data with the exception of the sub-theme of *'Small Pleasures'*. These data therefore also support all of the themes (except for *'Small Pleasures'*) identified in study 2. Ten new sub-themes were identified that could be incorporated into the existing themes. These sub-themes were: Symptoms of HD; Importance of Carers Health; Importance of Routine; Positive Aspects of HD; Access to Information; Ways of Coping; Positive Emotion; Treatment Issues; Religious Issues and Secrets in the Family. The sub-theme *'Small Pleasures'* was removed from the sub-

themes. All observed themes and sub-themes were integrated into the existing ComQoL-A5 (Cummins, 1997) to generate a HD specific QoL measure for spousal carers (HDQoL-C).

Method.

Sample:

47 participants took part in this study. The sample included 32 full time HD family carers (27 spousal carers and 5 'other' family members) and 15 part-time carers who also worked outside of the home. Although for the purpose of the study it was most relevant to obtain information from spousal carers, it was felt unethical to exclude other family members from the focus groups as they were conducted as part of a Huntington's Disease Association Family Day (2002). Moreover, although such family members may have different issues, their views in terms of caregiving were still deemed important. As HD is a rare condition and as such, there are only a small number of carers in the UK, some participants felt they would be inhibited from speaking frankly if they could be identified in any way. Therefore, to provide reassurance of anonymity to participants, no other sample characteristics were sought; however, the sample were predominantly white, British and female. All carers were recruited through the Huntington's Disease Association UK. All participants gave informed written consent and were advised of their right to withdraw from the study if they wished to do so.

Materials:

The themes from Study 1 (concept clarification) and Study 2 (Photovoice) were integrated and displayed on overhead transparencies in order that they could be raised effectively within a focus group setting, see Table 7.1 below.

Table 7.1. Integration of themes from Studies 1 & 2.

THEMES	SUB-THEMES
Levels of Support	Appropriate help from social services Professional knowledge and understanding Appropriate specialist services Support from health care professionals
Dissatisfaction with caregiving role	Duty of care Benefit of personal support Advocacy Burden of Responsibility Genetic Issues Loss of Identity
Practical aspects of caregiving	Appropriate care facilities Safety and Security Practical support Financial Burden Tiredness Lack of time Support from friends and family Daily Hassles
Feelings and emotional wellbeing	Loss of emotional closeness Isolation and Loneliness Negative emotions Future concerns Escape Sense of Loss Neglected Needs Small Pleasures

Facets from the three domains of *'Health', 'Productivity' and 'Place in the community'* were placed onto OHT's in order that they could be brought to the attention of the focus groups as it was unclear as to their importance to QoL in spousal caregiving from the data gathered in

study 1. Table 7.2 (below) contains these further items that were considered for discussion within the focus group setting.

Table 7.2 Items on the ComQoL-A5 considered for discussion at focus groups.

2(a)	How many times have you seen a doctor over the past 3 months?
2(b)	Do you have any disabilities or medical conditions? (e.g. visual, hearing, physical, health, etc.).
2(c)	What regular medication do you take *each day*?
3(a)	How many hours do you spend on the following *each week*? (Average over past 3 months). - Hours paid work, Hours formal education, Hours unpaid childcare.
3(b)	In your spare time, how often do you have nothing much to do?
3(c)	On *average*, how many hours TV do you watch each day?
6(a)	Below is a list of leisure activities. Indicate how often in an *average month* you attend or do each one for your enjoyment (not employment).
6(b)	Do you hold an *unpaid* position of responsibility in relation to any club, group, or Society?
6(c)	How often do people *outside your home* ask for your help or advice?

Finally, facets of the ComQol-A5 that had fallen into response scales 1 and 2 ('not at all important' and 'not very important') were placed onto OHT's in order that they could be brought forward for discussion at the focus groups. Table 7.3 below depicts these facet items.

Table 7.3. Facet Items considered 'not at all important' or 'not very important' to HD spousal carers QoL.

Q1 b)	How many personal possessions do you have compared to other people?
Q2 a)	How many times have you visited the doctor in the last 3 months?
Q2 c)	What regular medication do you take each day?
Q3 c)	On average, how many hours of TV do you watch a day?
Q6 b)	Do you hold an unpaid position of responsibility in relation to any club, group, or Society?
Q6 c)	How often do people outside your home ask for help and advice?
'Importance' Q1.	How important to you are the things you own?
'Satisfaction' Q1.	How satisfied are you with the things you own?

Procedure:

The focus groups took place consecutively at a Huntington's Disease Association Family Conference. Participants were initially introduced to each other and given a chance to become acquainted with the other members of the group. Participants were then given some background information about Studies 1 & 2, discussed tape recording the sessions and asked them to sign consent forms.

QoL was operationalised to the participants through Cummins' (1997) and the WHO (1995) definitions. However, carers were also advised that QoL was whatever they felt it meant to them as an individual. Examples of what might be a good quality of life and a poor quality of life were given in order to stimulate their ideas about QoL and to motivate them to think about themselves and participate in the groups.

Facets of the ComQol-A5 that had fallen into response scales 1 and 2 ('not at all important' and 'not very important') and objective vs subjective inconsistencies within the domains of 'Health', 'productivity' and 'place in the community' were presented on an overhead projector (OHP). Participants were then asked to talk about what each of these items meant to them in terms of QoL.

The integrated themes that had emerged from the qualitative data obtained in Study 1 (concept clarification) and Study 2 (photovoice) were again presented via OHP and participants were given the opportunity to discuss these newly generated themes in relation to QoL and spousal caregiving in HD.

The focus groups continued with participants being asked to talk about the main issues that enhanced or compromised QoL when caring for a spouse with HD. Finally, the researcher summarised the main points raised and asked whether participants' thoughts had been summed up adequately, whether anything had been missed and whether there was anything that should have been talked about that wasn't discussed. All focus groups were audio tape recorded and lasted between 1 and 1.5 hours.

Data Analysis:

Each focus group session was transcribed verbatim (see appendix VI, for transcript of Focus group 1). Therefore, the data represent 8.5 hours of transcribed material. Employing Interpretative Phenomenological Analysis (IPA, Smith et. al, 1995, Smith & Osbourn, 2004) the first transcript was read a number of times, the left hand margin being used to annotate what was interesting and significant about what the respondents said. Once this had been carried out for the whole of the first transcript, attention was returned to the beginning of the transcript and used the right hand margin to document any emerging themes. These themes were threaded back to the original transcript in order to validate their existence within the text. Emergent themes were then listed on a sheet of paper and studied for connections between them. All of the themes clustered together with ease to produce a number of superordinate concepts. As the clustering of themes emerged, they were continually checked in the transcript in order to make sure they worked for the primary source material. The next stage was to produce a coherently ordered table of themes in order to establish which themes most strongly captured the respondents' issues or concerns in

relation to their QoL. The clustered themes were given names that represented their overall superordinate theme and an identifier was added to each instance to aid the organisation of the analysis and facilitate checking back to the original transcript. During this process, themes were dropped if they did not fit well into the emerging structure or were not very rich in evidence. Themes from the first transcript were then used to ordinate the analysis of subsequent transcripts. As such, repeating patterns were established but the emergence of new issues was also recognised.

To determine reliability, an additional researcher undertook independent thematic analysis or the verbatim transcripts and intercoder reliability was established *(K =0.8)*. The identified themes were further validated through comparison with the domains of the ComQoL-A5 (Cummins, 1997). For anonymity, and also because there were a number of participants conversing on each tape (making accurate identification of participants difficult), names were replaced with *'M'* for male and *'F'* for female respectively. Furthermore, due to the amount of different participants conversing on each tape, it was difficult to accurately assess how many carers talked about each subject. Therefore, if it was clear how many carers talked about a particular issue, this was noted. Otherwise the term 'a number of carers' was used to denote a conversation between two or more carers.

Findings.

Emerging themes from the focus group data:

Participants were asked to describe in as much depth as possible the different ways that caring for an HD relative affected their overall quality of life. Participants' accounts clustered around four superordinate themes: Levels of Support; Dissatisfaction with Caregiving Role; Practical Aspects of Caregiving; Feelings and Emotional Wellbeing.

Levels of Support:

All of the carers in this study related how, as a consequence of caring for an individual with HD they had experienced difficulties in gaining access to the specialist services required and felt "let down by the system". Only four carers in this study reported a positive experience of receiving support from heath care professionals. The following carers' conversation captures much of the participants' disappointment with the support they receive from health care professionals and social services as well as the lack of knowledge and understanding that they often encountered. This is an interesting account as it also captures one of the carer's recognition of a helpful social worker who has tried to put some provision into place for this family.

M "....*These people who make the decisions obviously don't understand that they are messing with peoples' lives. Do they even know what HD is? They have never met Dad. The only one who*

has is the social worker who is as disgusted as us with the decision. I've just had Mum round here. She is over her initial upset and is now very angry with how the system has let us down. Bearing in mind we're in the UK, we aren't prepared to take this lying down and are prepared to fight and shout until we get the best for Dad. I don't want to see heads roll, I just think that a man who has worked all his life and is now in difficulties should be helped by the system he has paid so much into over many years. We are prepared to contribute to the care but they haven't even got back to us with any figures. Local MP, Downing street, newspapers, head of the social services".

F "I really am sorry this has happened to your family as I know what it is like to be told things like that. We fall through many cracks and I think I found too many in my searches for help. It got so that the word 'no' wouldn't phase me as I was so use to it. But if I ever hear a yes I do believe I would just pass out. I couldn't stand the excitement of it all. If I can help you in any way please let me know and keep us posted about your Dad as this is definitely not fair for him or your family".

The first carer to speak in this excerpt is expressing his annoyance with the system. He feels let down and angry at the way his father and their family have been treated.

"... .the system has let us down. Bearing in mind we're in the UK, we aren't prepared to take this lying down and are prepared to fight... "

He is motivated to do something to change the situation but is still in a state of mind where although he feels let down, he wants to resolve the situation rather than "see heads roll".

"I don't want to see heads roll, I just think that a man who has worked all his life and is now in difficulties should be helped by the system.. "

He still appears to be confident that they will win their fight for access to the appropriate services.

"..[we] *are prepared to fight and shout until we get the best for Dad".*

The second carer in this excerpt is offering her support to the first.

"I really am sorry this has happened to your family... "

However, she seems less positive in her attitude and almost resigned to the notion that as a HD carer, you have to get used to not receiving the professional support you require.

"It got so that the word 'no' wouldn't phase me as I was so use to it. But if I ever hear a yes I do believe I would just pass out".

She also suggests that the services are not actually available - that there is no one there to deal with all the issues that HD brings.

"We fall through many cracks and I think I found too many in my searches for help".

A number of other carers also felt that their role was compounded by a lack of appropriate specialist services for them to utilise. The following conversation describes how patients with HD often "fall through the net" as specialist services are simply not available to them.

F *"We don't fit in anywhere and for us as carers this is a difficult situation"*

M *"I totally agree"*

F *"As far as I'm concerned, if somebody needs respite as it were, it should be a health issue, they have HD and they should go to a specialised unit where they would get treatment while they are there"*

F *"Yeah I know, having HD is a major blow to any person. There are not enough, if any, proper health facilities that are able to handle HD patients. Most doctors know little about the disease. We have very few places to turn for help when the patient is still in the outside world and yet cannot care for themselves properly or refuse to take medications or otherwise place themselves in harm's way. Constant work by all of who are survivors must help make this change. Because until HD is either cured or arrested, there is a great need for facilities to handle people such as my brother. There are many HD patients wandering the streets and just considered insane... "*

The final carer in this excerpt especially highlights the lack of services available to cope with HD patients and a lack of knowledge from professionals.

"There are not enough, if any, proper health facilities that are able to handle HD patients. Most doctors know little about the disease."

She seems tired and overworked and there appears to be an element of despair in her comments that there is no one there to help her.

> *"We have very few places to turn for help when the patient is still in the outside world and yet cannot care for themselves properly"*

Dissatisfaction with caregiving role.

All of the carers related how, as a consequence of caring for a loved one with HD they had had to deal with a number of personal encumbrances. One pertinent issue was the 'duty of care' that many carers felt was placed upon them. The following account is from a carer who has tested positive for HD herself and shows how she feels a duty of care to her mother but is finding it increasingly difficult to cope with the burden of this responsibility whilst worrying about the genetic implications of HD herself.

> F *"I know that there must be others who have HD and also are the sole caregivers to a parent, but it feels like there is no place to go where I can express the pain involved in this and the guilt because there are times I feel as though I can't take it anymore. It is often a painful place mixed with cries of despair and then times when all goes as well as can be expected but it seems lately I am running out of the heart to go on being the one responsible for mum's needs while feeling the disease myself but there is no-one else to help, it is my job."*

This carer clearly expresses how she feels duty bound to care although she is not coping well with the situation and is "feeling the disease herself".

"I am running out of the heart to go on being the one responsible for mum's needs while feeling the disease myself but there is no-one else to help, it is my job".

Further, she expresses how this is a burden to her and seems to suggest that it takes 'heart' and possibly courage to care for someone with HD.

"I am running out of the heart to go on being the one responsible"

She also seems to be alone in her situation, she has no one to confide in, to express how she is really feeling. It is as if there is no one there to care for her.

"but it feels like there is no place to go where I can express the pain involved in this and the guilt because there are times I feel as though I can't take it anymore".

A number of other carers, touched on this issue further by discussing the notion of advocacy. They talked about how they felt as if they had lost their identity and that no one was advocating on their behalf.

M *"In that way it is difficult to get people to advocate for you, to stand up for you as a person and not just the partner of someone with HD"".*

Carers also emphasised the benefit of personal support from friends and family or from help groups. This kind of support seemed to differ from practical support and appeared to be a

source of strength for carers. The following account is of two of carers who are encouraging one another to find somewhere where she can find this kind of support for herself.

F *"When I was helping to care for my father, I remember how difficult it was I started going to one [a support group] it's small and I travel 150 mile round trip but it is so worth it. That would also give you a day too just you, take a little extra time and treat yourself to dinner or something. Take care of yourself first. You are no good to anyone else if you're falling apart".*

F *"Yes, you need to let off steam in a safe environment without feeling guilty about the person you are looking after. You know so you can voice your issues, our problems and not feel that you're letting anybody else down or its not important because the most important thing is the person with the disease and that why I feel that, and I think its come out for me today as well that um, that's where I think there is a lot to be done that isn't being done at all, to actually help carers to look after their own needs as well".*

These carers talk about how they have a need to talk about personal issues but they do not always feel at liberty to do so. They express a feeling of guilt as if by talking about all the difficulties they experience, as carers they are not being loyal to their affected loved one.

"you need to let off steam in a safe environment without feeling guilty about the person you are looking after. You know so you can voice your issues, your problems and not feel that you're letting anybody else down or it's not important"

The first carer recognises the benefits of such support and further suggests that having a bit of space to just think about herself for a while is extremely useful.

> *"That would also give you a day too just you, take a little extra time and treat yourself to dinner or something. Take care of yourself first. You are no good to anyone else if you're falling apart".*

However, a number of carers talked about different ways they had of coping.

> M *"The strain of coping with the HD and other responsibilities seems to keep knocking me back. I keep trying to get myself off the anti-depressants, but always end up taking them again when I feel I'm going downhill again. I have had some very good counseling, that has certainly helped"*

The above carer is clearly finding it difficult to cope with caring for his HD affected wife. He talks of being "knocked back" and "going downhill". However, he is being productive in going for counselling to help ease his situation.

Furthermore, a number of carers noted the benefits of having a religious faith to draw comfort from.

> F *"When I was helping to care for my father, I remember how difficult it was (sigh) especially looking and feeling like I was somehow seeing my own future played out before me. I'm glad the Lord God is your strength, I can relate. He is mine also. As difficult as it is sometime, remember that is a tremendous amount of strength".*

This carer gives words of comfort to another by encouraging her to remain strong.

"remember that is a tremendous amount of strength"

Practical aspects of caregiving.

All the carers also commented upon their struggle with the practical side of their caregiving role. A lack of appropriate care and respite facilities appeared to have a huge impact on many other practical issues such as safety, tiredness and lack of time. In the following account, this carer talks about the impact of HD on his life in terms of respite. He highlights the importance of appropriate care facilities and demonstrates how this impacts upon his time. He draws attention to the fact that he will have to live with this for the rest of the patient's life, thus indicating that caregiving in HD is both chronic and tiring.

M *"I think for me, it's the fact that the disease keeps evolving and changing. There is no time for a break, respite is difficult, as it can be hard to find and is not always suitable. You know you are going to have to live with this for the rest of their life".*

Another carer continues the conversation commenting upon the non-specialist care facilities that his father has been placed in.

M *"My Dad has been in the acute psychiatric admissions wing of the local hospital for 7 months now and was diagnosed with HD about 5 months ago. The ward he is in is not ideal as*

they have no specialist knowledge of HD and have themselves admitted they overlook him as he is quiet and doesn't bother anyone – apart from for his medication".

One carer talked about issues of safety with regards to her family member who is now in full-time care. She feels it is difficult to have her loved one home, as the accommodation is not suitable for their needs.

F *"I do not have HD but have cared for a family member who is now in full-time care. I have not been well with migraines this week but will make the effort Saturday or Sunday. She is happy and well cared for. It will be her 50th Birthday in a few weeks and I would love to have her home for at least one night. Our house is in disarray at the moment with kitchen renovations so I'll have to ensure it's safe first as she falls easily; it's not worth the risk if I can't ensure her safety".*

This carer is also justifying why she hasn't seen her family member for a while. She mentions she has not been well herself but suggests that she usually sees her regularly. The fact that this carer hasn't been able to visit her loved one this week seems to be concerning her. She seems to feel guilty both about the care situation and the lack of time that she has spent with her this week.

"I have not been well with migraines this week but will make the effort Saturday or Sunday. She is happy and well-cared for."

Access to information was another big issue for carers. The account below captures the struggle one carer has encountered in trying to access the correct information in order to adequately care for his daughter.

> M *"I find it difficult to find information, the information that I need and the information that I feel professionals should be able to give me! I struggled through years of searching for information which would help me in caring for my daughter at home. I know all too well the frustrations one can feel when faced with a caring need and not being able to find a resource to help you either solve it, understand it or where to go for help. Caregivers have enough responsibilities on their hand to spend hours researching for help".*

He talks about the frustrations of trying to care when you cannot find the correct resources.

> *"the frustrations one can feel when faced with a caring need and not being able to find a resource"*

Furthermore he sees the task of searching for information as an extra responsibility that he does not have time for.

> *"Caregivers have enough responsibilities on their hand to spend hours researching for help"*

Many carers also talked about how tired or exhausted they were from carrying out their caregiving role. This carer comments upon the exhaustion you encounter in a chronic caregiving situation.

F *"Being a full time caregiver is very stressful, you feel exhausted and need help to get out away from the house for a time so you can recharge".*

She also comments upon the need to get away to "recharge" and how you require help to do so. This is something that is especially pertinent in light of the previous comments raised about lack of help from appropriate specialist services.

"... .need help to get out away from the house for a time so you can recharge".

Some carers were given the "time-out" that they needed through practical support from friends and family.

F *"..trying to help the rest of the family cope with what to expect and how to help is way too much for one person, I have a strong family support system, and a group to help out".*

A lack of time combined with a high level of exhaustion led a number of carers to talk about the daily hassles that impact upon their lives. Daily chores such as washing or cooking the dinner had become hindrances for the carers as they tried to manage their caregiving role. The following account captures the way that small problems can be stressful for HD carers.

M *"And once you've planned your day and the cooking and the washing and whatever you are doing, the shopping, the gardening. The only thing that you do get wound up a little bit about is when something goes wrong. If the washing machine goes wrong, or the cooker or something goes wrong, with the house then that's when it all, that's when you get wound up a little bit".*

F *"but don't you find it difficult to cope with doing all these things?"*

M *"yes, yeah, when something goes wrong"*

F *"so you are dependent on everything running ok and it only takes one tiny thing to make everything fall apart. It's like being on a short fuse... "*

M *"[yeah , yeah"*

F *"... and when one little things goes wrong it can feel like the world is crashing down"*

The first carer to talk in this account demonstrates how as long as everything is going well, he is able to cope but that it just takes a little thing to go wrong and he is starting to feel stressed.

"The only thing that you do get wound up a little bit about is when something goes wrong".

The second carer is confirming this statement but is stronger in her view point.

"... and when one little things goes wrong it can feel like the world is crashing down."

Carers also talked about their health and how it was important for them to be in good health. Interestingly, they did not seem to be interested in their health from their own perspective but from the perspective of what would happen to their HD affected loved one if they were unable to care.

F "I want to try to make some progress on the acknouledgment of the illness. I feel I have nothing to lose inasmuch as I've tried pretending that nothing is wrong (for years and years) and that

has had a bad effect on my health. I know that if I'm to be of any use to my family in the long term I have to recognise and look after my own health too. Balancing all of the family's needs when HD is involved seems to be quite a tough thing".

This carer appears to be more concerned about the family's needs than her own and sees her own health as a way of continuing to care for them all adequately.

"I know that if I'm to be of any use to my family in the long term I have to recognise and look after my own health too."

One final theme that clustered into this category was the issue of financial burden. All of the carers in this study reported concerns over money and how they were going to manage. Such financial issues often had different causes, for example, worries about the financial burden of caring and financial implications of genetic testing.

F *".. the other part that goes with it is the costings. We won't have anything to do with it as the carers we, we make sure that - there is a real implication between what the government are defining as healthcare and social care and when you back to the finances cos they're all interlinked there are a number of hoops that you've already got with a neurological terminal condition, its ridiculous, there should be a much simpler way, um and I think a a a way of relief to come come is, I know genetics can't be sort of forced upon anyone but there's got to be a better way of one the diagnosis is there the doors automatically open, the hoops shouldn't have to be jumped through cos you're not gonna fiddle when you've got a terminal condition like that one".*

The above carer feels angry at the 'hoops' that have to be jumped through to get some financial help to care for an affected loved one.

F *"its ridiculous, there should be a much simpler way.."*

A young family member comments upon the way that needing to work has separated their family.

M *"Mum, who still needs to work and so we cannot have Dad home... "*

The following account is from a carer who is describing what happened when he first found out that one of his parents had been diagnosed with HD. He reports a feeling of wanting to know his own genetic status but the financial implications that this knowledge brings with it result in him changing his mind.

M *"I rushed off to my GP to arrange genetics counselling and testing. Between then and the appointment I was advised not to test, mortgage, life insurance are harder to get with a positive result, so I decided against it... "*

The account from the carer below is very similar to the previous carer's dialogue. This carer is also 'at-risk' of HD and is thinking of the disease in terms of the financial implications it may have for her in the future.

F *"I'm 20 and my Mum's just tested positive for HD, with a CAG repeat count of 44. Seeing as I have a 1 in 2 chance of developing it, is it wise for me to start up a pension fund as soon as possible since I'll not be able to work for as long as other people?"*

All of the carers in this study raised issues that were entirely specific to HD in terms of the nature of the disease itself. Carers highlighted a number of issues in relation to symptomology, routine, treatment and also some positive aspects of HD. The account below captures the importance of routine to the HD patient and subsequently the carer. Such routine is also bound up with a rigid mindset that is often evident in HD (Harper, 1996).

M *"She has to have her meals absolutely bang on the dot of time, breakfast at 8 'o' clock, coffee again this must be at the proper time, dinner at 1 'o' clock and tea must be at 6 'o' clock. Now, if something's gone wrong, I've got caught up doing something else, then I'm rushing and tearing about and I get a little bit upset (voice wavers) because I can't get the dinner cooked on time and she's upset cos that's the way they like it, they like their meals and different things every day at that particular time, well that's how X's (HD affected individual) mind works now, she must have.."*

F *"[but it didn't before?"*

M *"no it didn't worry her before but it does now you see, before things never used to worry her what time she had her meals or things like that but now it does that's the difference in your life. I've got all the garden and all the shopping and cooking and everything so so that all the work has to stick to a particular time. But I, I, I, I've worked it, it works for me ok now, I've got everything sorted."*

F *"[you do get used to it"*

F *"yeah you get used to it, yeah I've got used to, the only thing like I say that upsets it is when something goes wrong with your household"*

M *"you have to build up to approaching change with caution whereas um, you might think it would be nice to go out for a day, you need to feed in well ahead those thoughts and ideas for them to be able to accept them, for the sufferer to be able to take them on board so it's a two way negotiation over a period of time, not something you can do , you know straight away and you might not succeed and that can be a frustration as well."*

The first carer in this conversation is obviously upset by the strain that the patient's rigid mindset places upon him. He is trying his best to cope but struggling to maintain the daily routine due to the lack of flexibility of the patient.

"Now, if something's gone wrong, I've got caught up doing something else, then I'm rushing and tearing about and I get a little bit upset (voice wavers) because I can't get the dinner cooked on time and she's upset cos that the way they like it.."

The conversation continues with the carer noting that his wife's personality has changed due to the nature of HD.

"no it didn't worry her before but it does now you see, before things never used to worry her what time she had her meals or things like that but now it does that's the difference in your life."

He further goes on to describe how change has to happen over a period of time. He seems to show some upset and disappointment as he talks about wanting to go out and do things with his loved one but she is unable to deal with anything that breaks from their routine.

> *"you might think it would be nice to go out for a day, you need to feed in well ahead those thoughts and ideas for them to be able to accept them."*

A number of carers also talked about the symptomology of HD in terms of treatment issues. The following conversation really encapsulates a number of the symptoms of HD which make it so difficult for both the patient and the carer to cope. It further notes how "getting it right" can make all the difference in terms of life quality.

> F *"My mum has HD and has been in a nursing home for two years now. She can do nothing for herself. I visit her once a week. But it is getting harder. A few weeks ago when I was visiting her she knocked me off a chair, smacked me and kicked me .hh The bits of speech I can understand from her is that she hates me and tells me to go home. I really don't know how to react. I ask her if she knows who I am and she answers the right person. So she does know its me. I am so bruised on the outside and inside. It hurts me so much to think my mother is like this to me. My friends don't understand how emotionally damaging this is. I know she is sick. This is the kind of stuff no one wants to talk about the stuff no one can see that HD really does. I can handle the fact of the movement, the eating problems and everything else that goes with it but what scares me the most is this stuff and how if I do get it I never want to do this to my own children. This is part of the reason she is in the nursing home. Then there is that guilt...I am the oldest of four children, and we let our mother who has given her whole life to take care of us in there."*

F *"I sort of know what you're going through as my Dad was getting increasingly violent and aggressive before it all came to a head and we had to get Dad into hospital. Um, he was in an acute psychiatric ward for about 7 months and from there we got him into a care home where he's been since February. Only once was any violence aimed at me – usually Mum got the rough end of the stick - but it still shook me up a lot. Now though, several months down the line things are a bit more under control. Dad is on Oxazepam for the anxiety and Olanzapine which is a mild anti-psychotic. He still has no chorea to speak of but his balance is gradually going and he falls over a lot. A year ago, I thought our world was collapsing but now, with the right kind of care and the right mix of meds things are slowly looking better, never perfect just better than before."*

The first carer in this conversation highlights the anxiety she has experienced due to her mother's symptoms.

"I am so bruised on the outside and inside. It hurts me so much to think my mother is like this to me."

She has been subject to violence and abuse yet still feels guilty about placing her mother in a nursing home.

"Then there is that guilt...I am the oldest of four children, and we let our mother who has given her whole life to take care of us in there."

Moreover, the genetic implications of HD are again evident as she shows concern that she may behave the same way towards her own children.

" I can handle the fact of the movement, the eating problems and everything else that goes with it but what scares me the most is this stuff [violence] and how if I do get it I never want to do this to my own children."

The second carer in this conversation tells a more hopeful story. She too has experienced the violent outbursts often associated with HD.

"Only once was any violence aimed at me – usually Mum got the rough end of the stick - but it still shook me up a lot."

However, her father is now in a nursing home where with the right treatment, his behaviour has improved and life has become much easier to cope with.

"A year ago, I thought our world was collapsing but now, with the right kind of care and the right mix of meds things are slowly looking better never perfect just better than before."

Feelings and emotional wellbeing.

All of the carers related how, as a consequence of caring for a loved one with HD, they experienced a number of emotional issues that appeared to impact quite heavily upon their QoL. One of the most distressing emotional issues was an area that was only applicable to spousal carers, who described a sense of loss in terms of their marital relationship. The

account below shows how the carer is almost mourning the loss of his relationship with his wife. He acknowledges that is the nature of HD but is still finding it difficult to come to terms with the changing nature of his feelings.

M *"I have always enjoyed my wife's company, but am now having to build a more independent social life outside the home. I get these alternating feelings of sadness at the gradual passing of a warm and loving relationship with my wife, then anger at the behaviour. I think the worst thing is this roller-coaster nature of the mood swings so I never quite know who I'm going home to. I know this is normal for HD carers, but still hard to handle. I should add that I love my wife deeply and there are times when we still enjoy the warm relationship we always used to have. I know it's the illness, not her, that's to blame. That's another ramble on, but again it feels good to get it out in the open."*

Carers also felt isolated and lonely. In the account below, the carer talks about how her and her husband have actually isolated themselves from other people.

F *"we don't want anyone to tell us, we don't want anybody in the house, we'll cope with this on our own and its all the burden on all the carers then. Cos you have that burden that they don't want anybody in the house and they deny that they are ill."*

She also doesn't seem to want any involvement from outside parties.

"we don't want anyone to tell us, we don't want anybody in the house, we'll cope with this on our own"

However, this decision is clearly proving to be a burden to her.

> *"all the burden on all the carers"*

A number of other carers identified explicitly their need to escape from their caregiving role. The carer's account below identifies how emotionally wearing caregiving is for him and how he feels the need to 'run away' from it all.

> M *"When she is ok I almost think that there is nothing wrong and everything will be ok after all. Then she gets bad again and I just want to run a mile. It really wears me down not knowing which person I'm going to be with each day."*

All of the carers experienced a number of negative emotions such as depression, guilt, anger and frustration combined with a sense of loss in relation to their caregiving role. The account below encapsulates some of these negative emotions.

> F *"The other thing, the actual loss thing, its always there isn't it at the back of your mind. When you've talked to other groups did you pick that up, that its not always expressed? It is almost accepted that that you're dealing with a loss. The word that jumps to mind is hardened, it isn't hardened its more of a suppressed emotional feelings and guilt mixed with anger and I've picked that up with other people that we've talked to. I think you look at people how they might have been or how you were going to be with them. Does that make sense to you?"*

This carer seems to be addressing a number of different things within her account above. She is expressing a lot of negative emotions.

"suppressed emotional feelings and guilt mixed with anger"

She is also expressing a sense of loss and seems to be hurting from both her current situation and also the loss of a future.

" I think you look at people how they might have been or how you were going to be with them"

A number of carers also talked about their shock of finding out that HD was present in the family and felt distressed by the secrets that had been kept from them.

F *"We only found out that HD was in the family recently and it was such a shock. We are still having troubles getting information from any of the family, we still haven't spoken to my mother-in-law, and now my brother-in-law is acting the same, in the space of one conversation he told my husband that he had the test and was negative, and then told him that if my husbands test comes out negative, then he will get tested himself as he might be the other 50%. I don't understand why he would lie like that, especially in the same conversation".*

Carers also talked a little about their own needs and how these were often neglected. In the carer's account below, she talks about having to compromise her own needs in order to care for her loved one.

F *"you compromise, you compromise your own needs to make life easier and that compromise is ongoing. The other thing is that if you don't, aren't prepared to make that compromise, the sufferer then has increased anxiety which we now know worsens the condition."*

There were also some positive emotions that emerged from the carers' conversations. The account below shows a positive emotional response to HD from a carer who is 'at-risk' herself.

F *"I found out HD is in our family and I am at risk, my dad, my brother, but never for a second would I wish to never be born, life is the gift I want everyone to remember that what may seem like a curse now can really be a blessing."*

Within the conversations it also appears that discussing such emotional issues with like minded others has a therapeutic benefit to these carers. The carers have been given the opportunity to talk about very personal and difficult situations with people who can empathize with their feelings and experiences and they do so without prompting and express the release that unburdening themselves brings.

M *"it feels good to talk about my feelings... "*

This carer also talks about the positive aspects she feels that HD has brought her life. The following account demonstrates the way in which a family have become closer due to the genetic and devastating nature of HD.

F *"I have two boys of my own who are 15 and 12. I am in the early stages of the disease. I have only known this for about 6 weeks now but I've been at risk for the past 10 or 11 years. In these years I have talked to my boys about HD but of course they were very young at the time so I kept it very brief with them so that they wouldn't be afraid of my dad who has HD. Now that I know I have HD and that the boys are older I have been very open with both of them. They both know they're at risk, they know that mum has HD like grandad and our family will go through many changes. Also that our family will have to adjust to whatever mum is going through at the time some of it will be scary for them but just because mum has HD doesn't mean that either one of them will have it, but just like we teach our kids to protect themselves from diseases or from pregnancy they also have to know that this possibility is there and they need to make all the right decisions when they find that special someone. It's hard enough just being a young adult now a-days let alone having to worry about this horrible disease but we have no choice if we deny that the disease exists what good will that do we have to remain strong and open with our kids at all times so that they know we love them and support them and as a family we can get through what ever life's challenges are. HD has brought us closer."*

This carer looked after her father and is now living with HD herself. She talks about being open with her children and about adjusting family life in order to deal with the disease.

"Now that I know I have HD and that the boys are older I have been very open with both of them. They both know they're at risk they know that mum has HD like grandad and our family will go through many changes. Also that our family will have to adjust to whatever mum is going through."

She appears to be very philosophical about HD and talks about her children making the right life decisions in light of the fact that they are 'at-risk' of the disease.

> *"just like we teach our kids to protect themselves from diseases or from pregnancy they also have to know that this possibility is there and they need to make all the right decisions when they find that special someone."*

Finally, she acknowledges how HD has in some way benefited her family as it has brought them closer together.

> *"as a family we can get through whatever life's challenges are. HD has brought us closer."*

Specific Questions regarding the ComQoL-A5 Domains of *'Health'*, *'Productivity'* and *'place in the community'*

Facets from the three domains of *'Health'*, *'Productivity'* and *'Place in the community'* were also discussed within the focus groups in order to identify their importance to the QoL of HD spousal carers. The carers were asked to talk about how relevant these particular issues were to their QoL and try to give examples. It was decided that if the questions received only one favorable comment they would be included on the pilot of the HDQoL-C as they could be

removed after factor analysis if necessary later. Each Domain question is discussed in turn below and includes corresponding excerpts.

Health:

Question 2(a) How many times have you seen a doctor over the past 3 months?

When asked whether the above question was important to spousal carers, the participants in these focus groups predominantly felt that their actual health was more important than whether they had been to see the doctor in the past few months. They also commented that they needed to be healthy in order to care but that they did not consider their health that much if it was not interfering with their caregiving role.

F *"I don't really think about whether I am feeling well or not, only whether I am able to care for X (HD affected individual). I guess I am concerned about my health because if I get ill, who will look after X"*

Question 2(b) Do you have any disabilities or medical conditions? (e.g. visual, hearing, physical, health, etc.).

Carers also agreed that medical conditions could be a concern for them. However, they were again more concerned about whether disabilities or medical conditions would hinder their ability to care for their loved one.

F *"It's not something you really think about from day to day. It only matters when something like that stops you from being able to do all the things you need to do for X... "*

Question 2(c) What regular medication do you take each day?

Carers felt that this question was implicit within question 2(b) in that if they had a medical condition or disability, they would probably be taking medication for it.

M *" Well it stands to reason really, if you are ill, then you take whatever will get you back on your feet again as quickly as possible.."*

Productivity:

Question 3(a) How many hours do you spend on the following each week? (Average over past 3 months). (Hours paid work, Hours formal education, Hours unpaid childcare).

When questioned as to whether the above question was important to spousal carers, the participants in these focus groups predominantly felt it was difficult to identify how much

time they actually spent on any activity. They also felt that the question was not entirely relevant to them as it asks how many hours of a) paid work; b) formal education and c) unpaid childcare, formal education being something they felt they wouldn't have time for. The carers felt that this question would be more appropriate if question b) asked how many hours of unpaid caring they did.

M *"It is difficult to work out when I am doing things for the kids and when I am doing things for X. It can be a bit like having an extra child in the house. Well it is really, another person depending on you, you know.. "*

F *"I wouldn't get time to do any education type stuff.."*

F *"It [the question] doesn't ask how much time I spend caring for X."*

Question 3(b) In your spare time, how often do you have <u>nothing</u> much to do?

Initially the carers felt that this was an irrelevant question as they never had nothing much to do and very little spare time. However, when they were shown how this question could help to identify whether they have enough time for themselves, the carers agreed that this was (from that perspective) an important question. They felt that the words 'In your spare time' should be removed to make the question more relevant to them.

F *"I still say that we never have nothing much to do but I get what you mean. If we never have any time for ourselves then this is wrong.."*

M *"I think if you just asked if we have any spare time then the question would make more sense to us. Otherwise, it just makes me annoyed to think that anyone would think that I have any spare time where I do nothing. It's like the question has been made up by someone doesn't know what it its like to be a carer."*

Question 3(c) On average, how many hours TV do you watch each day?

The carers were unanimous in their decision that this was not a relevant question in relation to the HD spousal carers QoL. They suggested that watching TV was a pastime that they rarely took part in unless they were also doing something else at the same time, e.g. ironing or feeding their loved one. Although watching TV could be evidence of the carers having some relaxation time, the carers interpreted it as a question related to having a sedentary lifestyle.

M *"TV... Whats that!!"*

F *"If we are having a quiet day then I sometimes get a chance to sit down and watch a bit of telly with X (HD affected individual). Its never my choice though, he has his set programs and there's no changing the channel."*

M *"I really can't see what this question could tell you about my quality of life.."*

Place in the Community:

Question 6(a) Below is a list of leisure activities. Indicate how often in an average month you attend or do each one for your enjoyment (not employment).

When asked whether the above question was important to spousal carers, the participants in these focus groups predominantly felt that this question was more relevant to them in terms of time issues and neglected needs rather than in relation to their place within the community.

F *"I don't think about my place in the community, I think about my place in my family. That's where I belong and that's where I'm needed. I think this question just makes me think about some of the things I would really love to do for myself but never get a chance to.."*

Question 6(b)Do you hold an unpaid position of responsibility in relation to any club, group, or Society?

The carers were unanimous in their decision that this was not a relevant question in relation to HD spousal carers QoL. They felt that due to their caregiving role, such a position of responsibility would be something that they would never consider. They also felt that having such a position of responsibility would not benefit their QoL.

M *"This would be far too much of a commitment and I can only think of this as something really really stressful."*

Question 6(c) How often do people outside your home ask for your help or advice?

Once again, the carers were unanimous in their decision that this was not a relevant question in relation to HD spousal carers QoL. They felt that this was something that rarely happened. This was partly due to the fact that they were always incredibly busy and they felt that people who knew them would not overload them further with their own issues. Moreover, a number of the carers believed that they were often too wrapped up in their own problems and wouldn't necessarily notice even if they were asked for help or advice.

F *"People don't usually ask me for my help, I think they know what the answer would be (laughs) and to be honest, I'm not even sure that if someone did need me to do something for them I would even take it on board. My minds always elsewhere, thinking of the next thing I have to do or worrying about what still needs to be done."*

Specific Questions regarding the ComQoL-A5 questions deemed 'not at all important' or 'not very important' to HD spousal carers QoL in Study 1 (Concept Clarification).

The facets of the ComQol-A5 that had fallen into response scales 1 and 2 ('not at all important' and 'not very important') were also discussed within the focus groups in order to

identify their importance to the QoL of HD spousal carers. A number of these questions (Q2a; Q2b; Q3c; Q6b; Q6c;) had been previously addressed when discussing the domains of *'Health'*, *'Productivity'* and *'place in the community'*. As such, only three questions were considered in this part of the focus group. These were Q1 b) How many personal possessions do you have compared to other people?; *'Importance'* Q1. How important to you are the things you own?; *'Satisfaction'* Q1. How satisfied are you with the things you own?. Each question is discussed in turn below and includes corresponding excerpts.

Question 1 b) How many personal possessions do you have compared to other people?

The carers were unanimous in their decision that this was not a relevant question in relation to HD spousal carers QoL. The notion of material-well being or wealth was a concept that was insignificant to the carers in this study. They were keen to note that they wished for enough money to live on and a home that was suitable for the patients needs. However, they were not interested in making comparisons between themselves and other people in terms of personal possessions.

F *"you tend to see material things for what they are – stuff really, you can't take it with you. It would be nice to have enough money to buy the things that X (HD affected individual) needs without worrying. Its horrible when you get the point where you are thinking, please don't mess those trousers up – I can't afford another trip to the dry cleaners.."*

M *"All the material possessions in the world wouldn't give us what we want. Reprieve from this nasty disease and the chance to grow old together is what really counts. The thing that is so sad is that people who have their health don't even consider it and waste their time always wanting to buy more stuff. If X (HD affected individual) had her health then we would cherish it and realise how lucky we are."*

Question 1. on the 'Importance' scale. How important to you are the things you own?

As with the previous question, carers agreed that this was not a relevant question in relation to HD spousal carers QoL. They were not interested in the concept of ownership of possessions.

M *"As I've just said, this just isn't important to us, it's not what life is about."*

Question 2. On the 'Satisfaction' scale. How satisfied are you with the things you own?

Again, carers agreed that this was not a relevant question in relation to HD spousal carers QoL. They did not believe that the things they owned could bring them the type of satisfaction that would benefit their QoL.

> **M** *"This is really the same as importance isn't it? Having stuff won't make us satisfied in any way. It wouldn't make my life quality better, it wouldn't take away the curse of HD."*

Interpretation of Findings in relation to Study 1. (Concept Clarification) and Study 2. (Photovoice):

The analysis of the verbatim transcripts identified four different themes and 36 sub-themes illustrating the ways in which spousal caregiving in HD impacts on QoL. These were Levels of support; Dissatisfaction with caregiving role; Practical aspects of Caregiving and Feelings and Emotional Wellbeing.

These data supported the identification of the four themes in Study 1 (concept clarification) and further identified additional HD-specific sub-themes.

All the previously identified sub-themes from studies 1 and 2 were also evident in the focus group data with the exception of the sub-theme of *'Small Pleasures'*. These data therefore also support all of the themes (except for *'Small Pleasures'*) identified in Study 2 (Photovoice).

Ten new sub-themes were identified that could be incorporated into the existing themes. These sub-themes were: Symptoms of HD; Importance of Carers' Health; Importance of Routine; Positive Aspects of HD; Access to Information; Ways of coping; Positive Emotion; Treatment issues; Religious Issues and Secrets in the family. The sub-theme *'Small Pleasures'* was removed. The revised themes and their corresponding sub-themes are depicted in table 7.5 below.

Table 7.5. Integration of themes from studies 1, 2 & 3.

THEMES	SUB-THEMES
Levels of Support	Appropriate help from social services Professional knowledge and understanding Appropriate specialist services Appropriate care facilities Support from health care professionals
Dissatisfaction with caregiving role	Duty of care Benefit of personal support Advocacy Burden of Responsibility Genetic Issues Loss of Identity Ways of coping Religious beliefs Treatment issues
Practical aspects of caregiving	Safety and Security Practical support Financial Burden Tiredness & exhaustion Lack of time Support from friends and family Daily Hassles Access to Information Health Importance of routine Symptoms of HD
Feelings and emotional wellbeing	Loss of emotional closeness Isolation & Loneliness Negative emotions Concerns for the Future Escape Sense of Loss Neglected Needs Secrets in the family Positive emotions Positive aspects of HD

Conclusions:

These data provide much evidence that QoL is indeed compromised in many ways and is an issue for HD carers. The carers in this study often neglected their own needs as their caregiving role and the disease process took over their lives as well as the life of their HD affected spouse. Furthermore, the results show there are some similarities with carers of people with other types of dementia (e.g Dura et al, 1990) but that there is a need to consider HD independently due to the unique nature of the disease. The change of marital role that inevitably comes with caring for a spouse with dementia is often compounded in HD by extreme isolation. Moreover, the unaffected spouse has to take on board the fact that HD may also have been transmitted to any children and as such they may be placed in a position of caregiving for a number of decades.

In the following study, the above themes and sub-themes are integrated into the existing ComQoL-A5 (Cummins, 1997) for piloting. The aim is to develop a HD-specific quality of life measure for spousal carers of Huntington's Disease patients (HDQoL-C).

Chapter 8.

Validation of the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C)

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Background.

Research into the experience of the HD spousal carer has established that carers experience a number of unique obstacles within their caregiving role (e.g. Hans & Koeppen, 1980; Kessler, 1993). However, there is still a clear need to establish methodically the factors that impact upon the HD spousal carer's situation and ultimately their quality of life. As a result of the previous three exploratory studies, the aim of this pilot study was to validate the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) for use (initially) with spousal carers of HD patients.

Method.

Sample:

87 HD spousal carers (33 men, mean age 59.64 years, sd 12.72; and 54 women, mean age 57.24 years, sd 15.09) took part in the pilot study. All carers spent over 40 hours per week caring for their affected spouse and as such, caregiving was deemed to be their full-time occupation. Their educational background ranged from secondary school to post-graduate level and none of the carers had cared for another HD affected relative in the past. Participants were recruited through the Huntington's Disease Association UK. All participants gave informed written consent and were advised of their right to withdraw from the study if they wished to do so.

Materials:

Constructing the Huntington's Disease Quality of Life Battery for Carers HDQoL-C.

The HDQoL-C was developed using the existing domains and facets of the COMQOL-A5 (Cummins, 1997) a well documented and validated tool for measuring QoL with the general adult population. The structure of studies 1 to 3, which were carried out to investigate the underlying properties and meaning of QoL to family carers of HD patients, were adapted from Jirojanakul & Skevington (2000).

The HDQoL-C and the existing facets on the ComQol-A5.

Following on from Study 3 (focus group discussion), two questions were revised and seven questions were removed from the ComQoL-A5. These questions are shown in Table 8.1 below.

Table 8.1. Questions on the ComQoL-A5 that were revised or removed after focus groups discussion.

REVISED Questions:	Revised to:
3(b) In your spare time, how often do you have nothing much to do?	How often do you have any spare time?
3(a) How many hours do you spend on the following *each week*? (Average over past 3 months): Hours paid work, Hours formal education, Hours unpaid childcare.	How many hours do you spend on the following *each week*? (Average Over past 3 months): Hours paid work, Hours unpaid caring, Hours unpaid childcare.

REMOVED Questions:
3(c) On *average*, how many hours TV do you watch each day?
Q2(c) What regular medication do you take *each day*?
6(b) Do you hold an *unpaid* position of responsibility in relation to any club, group, or Society?
6(c) How often do people *outside your home* ask for your help or advice?
Q1 b) How many personal possessions do you have compared to other people?
'Importance' Q1. How important to you are the things you own?
'Satisfaction' Q1. How satisfied are you with the things you own?

Demographic and objective questions were adapted from the ComQoL-A5 (Cummins, 1997). New questions were generated through issues raised in focus group discussion. Objective issues were established as being relevant / good predictors of QoL through a review of existing QoL literature. Table 8.2 (below) notes each objective question and gives an example of its relationship to QoL.

Table 8.2. Objective Questions and their relationship to QoL as demonstrated by existing literature.

Objective Questions:	Evidence of importance from literature:
1. Demographic Information: a) What is your date of birth?	Blane et al, (2004)
b) What is your sex ? (please circle)	Eckermann (2000)
c) What is the highest education you received ?	Mercier et al, (1998)
d) What is your marital status?	Salokangas et al, (2001)
e) How long have you known of the presence of HD in your family?	Hakimian (2000) -
f) How long have you been caring for an HD affected family member? g) Are you the main carer for your HD affected family member? h) What relation to you is the HD affected family member you are caring for? i) Have you previously cared for any other HD affected family members?	Kessler (1993)
j) Do you have any children? • *if so*, what was is their genetic status?	Tyler & Harper (1983)
2. what is your personal or household (whichever is most relevant to you) gross annual income before tax?	Fernandez-Ballesteros et al, (2001).
3. Do you have any disabilities or medical conditions? (e.g. visual, hearing, physical, health, etc.).	Ford et al, (2001)
4. How many hours do you spend on the following *each week*? (Average over past 3 months) - hours paid work, hours unpaid childcare, hours unpaid caring for pHD.	Evandrou & Glaser (2004)
5. Is your home suitable / suitably adapted for your family's needs?	Addae-Dapaah & Khei Mie Wong (2001)
6. Below is a list of leisure activities. Indicate how often in an *average month* you attend, or take part in, each one for your enjoyment (not employment).	Wendel-Vos et al (2004)

The newly generated domains and facets of the HDQoL-C.

Also as a result of focus group discussion, 36 HD specific QoL facets subsumed in four Domains were identified. These domains were Levels of Support, Dissatisfaction with Ccaregiving Role, Practical Aspects of Caregiving and Feelings and Emotional wellbeing.

In constructing the HDQoL-C, the domain 'Levels of Support' was amalgamated with the domain 'Practical Aspects of Caregiving' to create the domain 'Aspects of caregiving'; the domain 'Dissatisfaction with Caregiving Role' was renamed 'Dissatisfaction with life' and the domain 'Feelings and Emotional Well Being' was renamed 'Feelings about living with HD'. The Domains and Facets of the HDQoL-C pilot are depicted in Table 8.3. below.

Table 8.3. Domains and Facets of the HDQoL-C at pilot.

THEMES	SUB-THEMES
Practical aspects of caregiving	Appropriate Help from Social Services Professional Knowledge and Understanding Appropriate Specialist Services Appropriate Care Facilities Support from Health Care Professionals Safety and Security Practical Support Financial Burden Tiredness & Exhaustion Lack of Time Support from Friends and Family Daily Hassles Access to Information Health Importance of Routine Symptoms of HD
Satisfaction with Life	Duty of Care Benefit of Personal Support Advocacy Burden of Responsibility Genetic Issues Loss of Identity Ways of Coping Religious Beliefs Treatment Issues
Feelings about living with HD	Loss of Emotional Closeness Isolation & Loneliness Negative Emotions Future Concerns Escape Sense of Loss Neglected Needs Secrets in the Family Positive Emotions Positive Aspects of HD

The domain 'Practical aspects of caregiving' comprised factors surrounding the HD carer role *per se*. The domain 'Satisfaction with Life' comprised subjective levels of satisfaction

with life quality. The domain 'Feelings about living with HD' comprised specific emotional issues surrounding HD and life quality.

New questions were generated for the facets to be included at pilot and are shown in Table 8.4 below.

Table 8.4. Newly generated questions for the HDQoL-C Pilot.

Objective Questions:
In your opinion, is your home suitable / suitably adapted for your families needs?
Practical Aspects of Caregiving:
Do you have difficulty coping with caring for someone with HD?
Please list any difficulties that you have (e.g. dealing with behaviour, physical problems, emotional problems etc.
How easy is it to get access to any information that you may need about HD or caregiving?
How important is it for you to maintain a regimented daily routine?
How often do you receive appropriate help from social services?
How often do you have access to professionals who have specialised knowledge of HD and understand its implications?
How often do you have access to appropriate specialist services?
How often do you receive support for yourself when you need it?
How much support are you given by health care professionals?
How often do people fight for your rights as a caregiver?
How often do the genetic implications of HD impact upon your caregiving role?
How often do you have access to appropriate care facilities?
How often do you receive any practical support that you need?
How often do you experience a conflict of interest between what you want and what your HD affected relative wants?
Are you safe and secure at home?
Satisfaction with Life:
How *satisfied are you* with the treatment that your HD affected relative receives?
Feelings about living with HD:
I *feel* betrayed
I *feel* loneliness
I *feel a* sense of loss
I *feel* guilty
I *feel* that the future is bleak
I *feel* financially disadvantaged
I *feel* deprived of a helpmate / partner
I *feel* isolated

Table 8.4 continued..
I *feel* strong
I *feel* hope
I *feel* supported by health care professionals
I *feel* frustrated
I *feel* a burden of responsibility
I *feel* that I have lost my identity
I *feel* exhausted
I *feel* supported by family and friends
I *feel* sad or depressed
I *feel* angry
I *feel* stressed
I *feel* emotionally drained
I *feel* worried about the genetic implications of HD
I *feel* the need to escape my caregiving role
I *feel* like there are not enough hours in the day
I *feel* like my own needs are not important
I *feel* comforted by the belief that one day there will be a cure for HD
In some ways I *feel* that HD has had a positive impact on my life
I *feel* comforted by religious beliefs
I *feel* that I can cope
I *feel* that HD has made me a stronger person.
I *feel* frustrated by the daily hassles of caregiving
I *feel* overwhelmed by my caregiving role
I *feel* that there are too many secrets in the family
I *feel* like I have assumed a duty of care forced onto me
I *feel* like I have resigned myself to a life of caregiving
I *feel* like I don't know who I am anymore

Constructing the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) and the domains of 'Importance' and 'Satisfaction'.

The Comprehensive Quality of Life Scale for Adults (ComQoL-A5) (Cummins, 1997) contains questions that ask how satisfied an individual is with a quality of life domain (e.g. How *satisfied* are you with your health?) and questions that assess the importance of the domain to the individual (e.g. how *important* is your health?). Thus, allowing for *'Satisfaction'* and *'Importance'* to be multiplied in order to generate a QoL score for each domain based on how satisfied the individual is within a domain and further, how important it is to them.

However, more recent research by Cummins (2003) has demonstrated a strong positive correlation between the domains of *'Importance'* and *'Satisfaction'* on the ComQoL-A5. This means that *'Importance'* and *'Satisfaction'* are so closely related, that there is really no need for them both to be used within the same questionnaire. Therefore, in the construction of the HDQoL -C, the *'Importance'* rating will be disregarded and only a measure of *'Satifaction'* is included in the questionnaire.

Constructing the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) and subjective vs objective QoL.

Cummins also highlights the differences between subjective and objective QoL in his revision of the ComQoL-A5 (The Personal Wellbeing Index; PWI, 2004) which includes only subjective questions. Objective and subjective wellbeing are often not closely related in research (Cummins, 1997), for example physical health and perceived health are often poorly correlated (Rapley, 2003). This may be because objective measures quantify the items that are considered to impact upon QoL whereas, subjective measures reflect the individual's interpretation of a situation and its impact on QoL. However, objective QoL is still important to measure as it gives health care professionals a clear and tangible indicator of QoL which can be used in policy making and intervention research. Moreover, objective QoL can be assessed alongside subjective QoL to provide a richer description of the issues and concerns surrounding QoL for any individual.

Therefore, for the purpose of the HDQoL-C, demographic and objective data will be recorded independently to subjective QoL. It is anticipated that the subjective measure of QoL will give the researcher, or health care professional, an understanding of an individual's

perceived QoL which is highly salient in terms of psychological wellbeing. Additionally, as health care providers are increasingly asked to allocate their resources on the basis of outcome evidence, demographic and objective data can be utilized as a more tangible confirmation of the individual's subjective assessment.

The Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) at Pilot.

The pilot version of the HDQoL-C therefore comprised 63 questions, 7 demographic / objective questions and 56 subjective and HD-Specific QoL questions with at least one question to represent each facet (see appendix VII for HDQoL-C pilot version).

Procedure:

Spousal carers were given a questionnaire booklet that contained the pilot version of the HDQoL-C, the World Health Organization Quality of Life Short Form WHOQOL-BREF (WHO, 1996) (see appendix, VIII), the Satisfaction with Life Scale (SWLS) (Diener et al, 1985) (see appendix IX) and the Perceived Health Status Visual Analogue Scale (VAS, Weinman et al, 1995) (see appendix X).

The use of the three additional scales (WHOQOL-BREF, SWLS and VAS) allowed for data to be gathered in relation to congruent validity. Moreover, 10 carers answered a retest questionnaire after a two-week interval to gather data on test-retest reliability (Streiner and Norman, 1995).

Carers were asked to complete questionnaires independently and to reflect on life in the last three months. In order to assess face validity, participants were also asked to comment on the clarity of questions and words in the questionnaire. The amount of time taken to complete the questionnaire was recorded by the carers and any additional comments that had been written on the questionnaires were noted.

Findings:

Item analysis:

It was decided to employ Principal Components Analysis (PCA) in order to analyse the reliability and validity of the scale. However, there are some documented concerns with regards to the use of Factor Analysis in the development of QoL scales. For example, Juniper et al (1997) point out that as in factor analysis, items that are highly correlated are grouped together and items that have no strong correlation with any emerging domains and factors will be excluded from the questionnaire. This may lead to the exclusion of questions that are actually important to the population being studied. Furthermore Fayers & Machin (2001) note that although factor analysis is a powerful technique for exploring items during scale validation, it can be problematic in QoL research due to its causal nature i.e. there may not always be a causal link between a causal indicator and life quality but, an effect indicator may mediate between cause and QoL. For example a carer may be deemed to have a poor QoL due to their caregiving role, however it may be the nature of the relationship between

the carer and the care recipient that actually determines or mediates the impact of caregiving on QoL.

However, factor analysis has been used by a number of researchers to determine the final items to be included in QoL scales (e.g Bergner, 1993; Cummins, 1997; Hunt et al, 1986). Such scales are routinely used today and PCA is considered to be a valid technique to use in questionnaire validation (Fayers & Machin,2001). The term factor analysis is often used as an umbrella term for a variety of different techniques. One of the main distinctions is between principal component analysis (PCA) and factor analysis (FA). PCA is based on a transformation of the data set into a smaller set of linear combinations with all of the variance in the variables being used. However, in FA factors are estimated using a mathematical model. Both approaches produce similar results and are often used interchangeably by researchers (Pallant, 2003). However, Stevens (1996) argues that PCA is a psychometrically sound procedure that is simpler mathematically and avoids some of the potential problems with 'factor indeterminacy' that are associated with FA.

As such, PCA was used to explore the inter-relationship between the variables on the HDQoL-C and further, to refine and reduce the subscales of the HDQoL-C into a concise, valid and reliable scale that can be used to measure the QoL of spousal carers of HD patients. There are two main issues that need to be considered when employing PCA in scale development, firstly the sample size and secondly the strength of the relationship between the items on the scale. There is a non consensus as to how large the sample size needs to be for PCA, with some researchers suggesting at least 300 participants (Tabachnick & Fidell, 1996) and others arguing that it is not the sample size but the ratio of participants

to items that is important (Nunnally, 1978, cited in Pallant 2003). However, Stevens (1996), notes that the sample size requirements have been reducing over the years as more research has been conducted.

The sample size in this study is small with only 87 spousal carers completing the pilot questionnaire. However, because HD is such a rare disorder (prevalence rates being 10 per 100,000, Quarrell, 1999), the sample size is likely to be representative of the HD spousal carer population in the UK.

The second issue concerning the employment of PCA, regards the strengths of the inter-correlations between the scale items. Tabachnick & Fidell (1996) suggest that the correlation matrix should be inspected for coefficients greater than .3 and that if there are only a few found, then the data may not be suitable for PCA. Inspection of the items on the HDQoL-C pilot data set revealed many items showing correlations of .3 and above, thus suggesting the data was indeed suitable for PCA. Bartlett's test of sphericity (Bartlett, 1954) and the Kaiser-Meyer-Oklin (KMO) measure of sampling adequacy were also employed to assess the suitability of the data for PCA. Bartlett's test of sphericity was significant suggesting that the data was suitable for PCA. However, in the initial stages of PCA, a number of the items on the HDQoL-C were correlating highly thus providing evidence of multicollinearity. In each of these instances, one of the highly correlating questions was removed until the scale achieved an acceptable Kaiser-Meyer-Oklin value of .6 or above.

Principal Components Analysis (PCA)

Component 1 of the HDQoL-C was not subjected to PCA as this component requests demographic and objective information from the carer and each question is treated independently. Objective questions were generated through issues raised in focus group discussion and were established as being relevant and good predictors of QoL through a review of existing QoL literature (see Table 2, pg 158). The information in this section can be used in research to investigate the factors that may predict QoL in caregiving. Alternatively, it may be used by the practitioner to build up an overall picture of a carer. As this component does not in itself constitute a scale, the researcher / practitioner is able to omit questions or include additional questions that may be of interest in this section.

Components 2, 3 and 4 all comprise differing aspects of disease-specific and subjective QoL. However, they are worded in different ways and as such 'tap into' differing subjective aspects of life quality that cannot be incorporated into a single scale. That is, questions that ask *How often?* cannot be placed with questions that ask about *How do you feel?* as they are different forms of items. As such, these three components were subjected to PCA independently of each other to produce a battery of scales with which to assess the QoL of spousal carers of HD patients.

Component 2: Practical aspects of caregiving.

After addressing multicollinearity, the nine remaining items of component two were subjected to PCA using SPSS. Prior to performing PCA the suitability of data for factor analysis was assessed. Inspection of the correlation matrix revealed the presence of many coefficients of 0.3 and above. The Kaiser-Meyer-Oklin value was 0.70, exceeding the recommended value of 0.6 (Kaiser, 1970, 1974) and the Bartlett's Test of Sphericity (Bartlett, 1954) reached statistical significance, supporting the factorability of the correlation matrix.

Principal components analysis revealed the presence of three subscale components with eigenvalues exceeding 1, explaining 40.8 per cent, 17.81 per cent and 12.97 per cent of the variance respectively. Varimax rotation was performed. The rotated solution revealed the presence of simple structure (Thurstone, 1947), with each component showing strong loadings, and all variables loading highly on to only one component. The interpretation of the three components found 'levels of support' and 'access to professionals' loading strongly onto component 1, 'long term and genetic issues' loading strongly on to component 2 and 'daily hassles' loading strongly onto component 3. Table 8.5 below outlines the factor loadings for component 2.

Table 8.5. Factor Loadings for Component 2: Practical Aspects of Caregiving.

Questions:	Component		
	1	2	3
How much support are you given by Health Care Professionals?	.791		
How often do you receive any practical support you need?	.786		
How often do you receive appropriate help from social services?	.744		
How often do you have access to professionals who have specialist knowledge of HD and understand its implications?	.723		
How often do you sleep well?	.596		
How do the genetic implications of HD impact upon your caring role?		.907	
How often do you have access to appropriate care facilities?		.663	
How often are you restricted by the need to maintain a regimented daily routine?			.856
How often is there a conflict of interest between what you want and what you HD affected relative wants?			.605

Component 3: Satisfaction with life.

The 8 items of component 3 were subjected to PCA using SPSS. Prior to performing PCA the suitability of data for factor analysis was assessed. Inspection of the correlation matrix revealed the presence of many coefficients of 0.3 and above. The Kaiser-Meyer-Oklin value was 0.72, exceeding the recommended value of 0.6 (Kaiser, 1970, 1974) and the Bartlett's Test of Sphericity (Bartlett, 1954) reached statistical significance, supporting the factorability of the correlation matrix.

Principal components analysis revealed the presence of two subscale components with eigenvalues exceeding 1, explaining 46.33 per cent and 14.94 per cent of the variance respectively. Varimax rotation was performed. The rotated solution revealed the presence of simple structure (Thurstone, 1947), with each component showing strong loadings, and all variables loading highly on to only one component. The interpretation of the two components found 'overall quality of life issues' loading strongly onto component 1, and

'personal issues' loading strongly onto component 2. Table 8.6 below outlines the factor loadings for component 3.

Table 8.6. Factor Loadings for Component 3: Satisfaction with Life.

Questions:	Component	
	1	2
How satisfied are you with your overall quality of life?	.884	
How satisfied are you with your own happiness?	.878	
How satisfied are you with the treatment that your HD affected relative receives?	.752	
How satisfied are you with what you achieve in life?	.558	
How satisfied are you with feeling a part of your community?		.827
How satisfied are you with your close relationships?		.685
How satisfied are you with your health?		.654
How satisfied are you with how safe you feel?		.549

Component 4: Feelings about Living with HD.

After addressing multicollinearity, the remaining 17 items of component 4 were subjected to PCA using SPSS. Prior to performing PCA the suitability of data for factor analysis was assessed. Inspection of the correlation matrix revealed the presence of many coefficients of 0.3 and above. The Kaiser-Meyer-Oklin value was 0.71, exceeding the recommended value of 0.6 (Kaiser, 1970, 1974) and the Bartlett's Test of Sphericity (Bartlett, 1954) reached statistical significance, supporting the factorability of the correlation matrix.

Principal components analysis revealed the presence of two subscale components with eigenvalues exceeding 1, explaining 38.22 per cent and 14.15 per cent of the variance respectively. Varimax rotation was performed. The rotated solution revealed the presence of simple structure (Thurstone, 1947), with each component showing strong loadings, and

all variables loading highly on to only one component. The interpretation of the two components found 'negative feelings about life' loading strongly onto component 1, and 'positive feelings about life' loading strongly onto component 2. Table 8.7 below outlines the factor loadings for component 4.

Table 8.7. Factor Loadings for Component 4: Feelings about Living with HD.

Questions:	Component	
	1	2
I feel exhausted	.828	
I feel stressed	.798	
I feel isolated	.782	
I feel guilty	.727	
I feel financially disadvantaged	.711	
I feel like my own needs are not important to others	.697	
I feel like I don't know who I am anymore	.682	
I feel sad or depressed	.668	
I feel that I have had a 'duty of care' forced upon me	.532	
I feel worried about the genetic implications of HD	.458	
I feel that HD has made me a stronger person		.820
I feel hope		.698
I feel that I can cope		.689
I feel comforted by the belief that one day there will be a cure for HD		.655
I feel comforted by religious beliefs		.628
I feel that HD has had a positive impact on my life		.576
I feel supported by family and friends		.303

Therefore, PCA revealed the presence of seven subscales subsumed within the three components of 'Practical Aspects of Caregiving', 'Satisfaction with Life' and 'Feelings about living with HD'. The component 'Practical Aspects of Caregiving' therefore consists of three subscales identified as 'Levels of support and access to professionals', 'Long term and genetic issues' and 'Daily hassles'. The component 'Satisfaction with Life' therefore consists of two subscales identified as 'Overall quality of life issues' and 'Personal issues'. Finally the component 'Feelings about living with HD' therefore consists of two subscales identified as

'Negative feelings' and 'Positive feelings'. Each of these three components (or domains) and their subscales (of facets) were then subjected to further analysis in order to identify the reliability and validity of the HDQoL-C

Reliability and Validity of the HDQoL-C.

The data from the 87 carers were examined for floor and ceiling effects. Furthermore, the scale was assessed for face validity, content validity, congruent validity, internal consistency and test-retest reliability.

Distribution of answers:

Distribution of answers was examined for floor and ceiling effects (Ware & Keller, 1996). No item was found to show these effects, although the distribution of answers on some items was mildly skewed towards the lower end of the scales suggesting that respondents generally perceived that their QoL was poor in those aspects.

Validity:

The validity of a scale refers to the degree to which the scale measures what it is supposed to be measuring. Therefore, carrying out validity assessments on the HDQoL-C addresses the question of whether the scale does indeed measure the impact of HD on the QoL of

spousal carers. Face validity was investigated in order to establish whether the items on the scale seemed appropriate at face value. Content validity was investigated through the detailed examination of the questionnaire by a number of experts. Finally, congruent validity was investigated by correlating the HDQoL-C with a previously established QoL scale (WHO group, 1996) , a Satisfaction with Life Scale (Diener et al, 1985) and a Perceived Health Scale (Weinman et al, 1995) in order to provide evidence that it was able to 'tap into' the same construct as an already established and well-used QoL measure.

Face validity:

Carers were asked to read and comment on difficult or unclear terms. Question number 16, *"How often do people fight for your rights as a caregiver"* was seen as slightly confusing to the carers. However, this item was removed during factor analysis and no longer included in the questionnaire. Question 32 *" I feel that there are too many secrets in the family"* was considered irrelevant as although the carers did feel that at some point there had been too many secrets in the family, this was no longer the case. As such, this item was removed from the HDQoL-C. Carers also noted that having children impacts upon the caregiving role especially in consideration of the genetic nature of HD. As such, question 1j) was re-written: Do you have any children? *if so*, what was is their genetic status? (e.g. HD positive/ negative, 'at-risk'). Carers also requested a space on the questionnaire to write comments of their own.

Content validity:

Two experts in the field of QoL and two experts in the field of HD were asked to comment on the item content during the piloting of the HDQoL-C. In line with these comments, question 4(c) *"How many hours of unpaid caring do you do each week?"* was changed to *"How many*

hours of unpaid caring for your pHD (person with HD) do you do each week?" . Experts also suggested changes to a further three questions, however, these were removed from the questionnaire during factor analysis and as such, re-writing was unnecessary. One HD expert also noted that asking a carer to complete the HDQoL-C might actually give them the opportunity to speak to the carer on their own rather than in the presence of the patient. It was suggested that the mere existence of a questionnaire such as the HDQoL-C may give professionals a valid reason for asking to see the carer on their own without rousing the patient's suspicions about the nature of a private conversation between the carer and health care professional. Experts further noted that as HD can be so different from individual to individual, the carers' experiences were also likely to be widely varied. Experts also suggested a space on the questionnaire for carers to add any additional comments that they may have.

Congruent Validity:

All 87 spousal carers filled in a copy of the WHOQOL-BREF (WHO, 1996), a well documented and validated QoL measurement for use with the general adult population alongside the HDQoL-C in order to establish the validity of the new test. The relationship between the scores on the HDQoL-C and the WHOQOL-BREF revealed a Pearson's correlation coefficient of $r(86)=0.58$, $p \leq 0.01$ for component 2 (Practical aspects of caregiving), $r(86)=0.64$, $p \leq 0.01$ for component 3 (satisfaction with life) and $r(86)=0.76$, $p \leq 0.01$ for component 3 (Feelings about Living with HD).

Each component further correlates with the other two components at $p \leq 0.01$ as depicted by Table 8.8 below:

Table 8.8. Correlations Between the 3 subjective components of the HDQoL-C.

	Component 2: Practical Aspects of Caregiving	Component 3: Satisfaction with Life	Component 4: Feelings about Living with HD
Component 2: Practical Aspects of Caregiving	1.00	0.56	0.51
Component 3: Satisfaction with Life	0.56	1.00	0.83
Component 4: Feelings about Living with HD	0.51	0.83	1.00

Note:

Component 1 is demographical / objective data and each question is treated independently. Therefore, this component cannot be correlated with either the WHO-BREF or components 2,3, and 4 of the HDQoL-C.

In order to further assess the congruent validity of the HDQoL-C, each of the seven rotated factors (or subscales) that were established through PCA were individually correlated with the WHOQOL-BREF. This allowed for more in-depth analysis of the relationship between each of the individual subscales and QoL.

Component 2 of the HDQoL-C comprises three rotated factors (or subscales) relevant to practical aspects of caregiving. Subscale 1 (Levels of support and access to professionals) includes five questions: 'How much support are you given by Health Care Professionals?'; 'How often do you receive any practical support you need?'; 'How often do you receive appropriate help from social services?'; 'How often do you have access to professionals who have specialist knowledge of HD and understand its implications?' and 'How often do you sleep well?. The relationship between this factor and the WHOQOL-BREF revealed a

Pearson's correlation coefficient of $r(86)=0.509$, $p \leq 0.01$. Therefore, there is a significant moderate positive correlation between this rotated factor of the component 'practical aspects of caregiving' and the WHOQOL-BREF. Subscale 2 (Long term and genetic issues) includes two questions: 'How do the genetic implications of HD impact upon your caring role?' and 'How often do you have access to appropriate care facilities?'. The relationship between this factor and the WHOQOL-BREF revealed a Pearson's correlation coefficient of $r(86)=0.48$, $p \leq 0.01$. Therefore, there is a significant moderate positive correlation between this rotated factor of the component 'practical aspects of caregiving' and the WHOQOL-BREF. Subscale 3 (Daily hassles) includes two questions: 'How often are you restricted by the need to maintain a regimented daily routine?' and 'How often is there a conflict of interest between what you want and what you HD affected relative wants?'. The relationship between this factor and the WHOQOL-BREF revealed a Pearson's correlation coefficient of $r(86)=0.315$, $p \leq 0.01$. Therefore, there is a significant weak to moderate positive correlation between this rotated factor of the component 'practical aspects of caregiving' and the WHOQOL-BREF.

Component 3 of the HDQoL-C comprises two rotated factors (or subscales) relevant to satisfaction with life. Subscale 1 (Overall quality of life issues) includes four questions: 'How satisfied are you with your overall quality of life?'; 'How satisfied are you with your own happiness?'; 'How satisfied are you with the treatment that your HD affected relative receives?' and 'How satisfied are you with what you achieve in life?. The relationship between this factor and the WHOQOL-BREF revealed a Pearson's correlation coefficient of $r(86)=0.697$, $p \leq 0.01$. Therefore, there is a significant moderate to strong positive correlation between this rotated factor of the component 'satisfaction with life' and the

WHOQOL-BREF. Subscale 2 (Personal issues) includes four questions: 'How satisfied are you with feeling a part of your community?'; 'How satisfied are you with your close relationships?'; 'How satisfied are you with your health?' and 'How satisfied are you with how safe you feel?'. The relationship between this factor and the WHOQOL-BREF revealed a Pearson's correlation coefficient of $r(86)=0.372$, $p \leq 0.01$. Therefore, there is a significant low to moderate positive correlation between this rotated factor of the component 'satisfaction with life' and the WHOQOL-BREF.

Component 4 of the HDQoL-C comprises two rotated factors (or subscales) relevant to feelings about living with HD. Subscale 1 (Negative Feelings) includes ten questions: 'I feel exhausted'; 'I feel stressed'; 'I feel insolated'; 'I feel guilty'; 'I feel financially disadvantaged'; 'I feel like my own needs are not important to others'; 'I feel like I don't know who I am anymore'; 'I feel sad or depressed'; 'I feel that I have had a 'duty of care' forced upon me' and 'I feel worried about the genetic implications of HD'. The relationship between this factor and the WHOQOL-BREF revealed a Pearson's correlation coefficient of $r(86)=0.807$, $p \leq 0.01$. Therefore, there is a significant and strong positive correlation between this rotated factor of the component 'feelings about living with HD' and the WHOQOL-BREF. Subscale 2 (Positive feelings) includes seven questions: 'I feel that HD has made me a stronger person'; 'I feel hope'; 'I feel that I can cope'; 'I feel comforted by the belief that one day there will be a cure for HD'; 'I feel comforted by religious beliefs'; 'I feel that HD has had a positive impact on my life' and 'I feel supported by family and friends'. The relationship between this factor and the WHOQOL-BREF revealed a Pearson's correlation coefficient of $r(86)=0.444$, $p \leq 0.01$. Therefore, there is a significant moderate positive

correlation between this rotated factor of the component 'feelings about living with HD' and the WHOQOL-BREF.

The above in-depth analysis of the rotated factors of the HDQoL-C demonstrates that the sub-scales of the questionnaire positively correlate with an existing reliable and valid measure of QoL. Therefore, each of the sub-components of the HDQoL-C is measuring QoL as measured by the WHOQOL-BREF. Thus, providing further support for the HDQoL-C as a measure of QoL in HD spousal caregiving. However, it can be seen from the analysis that some of the factors correlate more strongly with the WHOQOL-BREF than others. This will be discussed within the context of the existing QoL literature in the following chapter.

All 87 spousal carers also filled in a copy of the Satisfaction with Life Scale (SWLS) Diener et al, 1985) which is a measure of the judgemental component, life satisfaction and is a five-item, self-report scale in which respondents rate their agreement on a seven-point likert scale with each item. The relationship between the scores on the HDQoL-C and the SWLS revealed a Pearson's correlation coefficient of $r(86)=0.295$, $p \geq 0.01$ for component 2 (Practical aspects of caregiving), $r(86)=0.425$, $p \leq 0.01$ for component 3 (satisfaction with life and $r(86)=0.449$, $p \leq 0.01$ for component 3 (Feelings about Living with HD).

There is therefore, a weak positive correlation between the SWLS and Component 2 'Practical aspects of Caregiving'; a significant moderate positive correlation between the SWLS and Component 3 'Satisfaction with Life' and a significant moderate positive correlation between the SWLS and Component 4 'Feelings about Living with HD'.

In order to further assess the relationship between satisfaction with life and the HDQoL-C, each of the seven rotated factors (or subscales) subsumed within the above three components, were individually correlated with the SWLS. This allowed for more in-depth analysis of the relationship between each of the individual subscales and satisfaction with life.

Component 2 of the HDQoL-C comprises three rotated factors (or subscales) relevant to practical aspects of caregiving. The relationship between Subscale 1 (Levels of support and access to professionals) and the SWLS revealed a Pearson's correlation coefficient of $r(86)=0.446$, $p \leq 0.01$. Therefore, there is a moderate positive correlation between this rotated factor of the component 'practical aspects of caregiving' and the SWLS. The relationship between Subscale 2 (Long term and genetic issues) and the SWLS revealed a Pearson's correlation coefficient of $r(86)=0.14$, $p \geq 0.05$. Therefore, there is a non-significant relationship between this rotated factor of the component 'practical aspects of caregiving' and the SWLS. The relationship between subscale 3 (Daily hassles) and the SWLS revealed a Pearson's correlation coefficient of $r(86)=0..325$, $p \leq 0.01$. Therefore, there is a weak positive correlation between this rotated factor of the component 'practical aspects of caregiving' and the SWLS.

Component 3 of the HDQoL-C comprises two rotated factors (or subscales) relevant to satisfaction with life. The relationship between Subscale 1 (Overall quality of life issues) and the SWLS revealed a Pearson's correlation coefficient of $r(86)=0.672$, $p \leq 0.01$. Therefore, there is a moderate to strong positive correlation between this rotated factor of the

component 'Satisfaction with life' and the SWLS. The relationship between Subscale 2 (Personal issues) and the SWLS revealed a Pearson's correlation coefficient of $r(86)=0.401$, $p \leq 0.01$. Therefore, there is a moderate positive correlation between this rotated factor of the component 'Satisfaction with life' and the SWLS.

Component 4 of the HDQoL-C comprises two rotated factors (or subscales) relevant to feelings about living with HD. The relationship between subscale 1 (Negative feelings) and the SWLS revealed a Pearson's correlation coefficient of $r(86)=0.535$, $p \leq 0.01$. Therefore, there is a moderate positive correlation between this rotated factor of the component 'Feelings about living with HD' and the SWLS. The relationship between Subscale 2 (Positive feelings) and the SWLS revealed a Pearson's correlation coefficient of $r(86)=0.618$, $p \leq 0.01$. Therefore, there is a moderate to strong correlation between this rotated factor of the component 'Feelings about living with HD' and the SWLS.

The above in-depth analysis of the rotated factors of the HDQoL-C further demonstrates a weak positive correlation between the SWLS and Component 2 'Practical aspects of Caregiving'; a significant moderate positive correlation between the SWLS and Component 3 'Satisfaction with Life' and a significant moderate positive correlation between the SWLS and Component 4 'Feelings about Living with HD'. However, by looking at the subscales individually, we are able to establish specific factors that have a non-significant relationship with the SWLS. Therefore, those items can be investigated more thoroughly in order to establish their relevance to the scale. For example, although all of the subscales of component 2 demonstrate only a weak relationship with the SWLS, factor 2 (Long term and genetic issues) is shown to have a non-significant relationship with satisfaction with life. It

would therefore be interesting to further examine the relationship between Long term and genetic issues and satisfaction with life in order to more fully understand the implications of this finding.

All 87 spousal carers also filled in a visual analogue scale (VAS) of perceived health status, a simple and practical way of obtaining a global measure of health (see Weinman et al, 1995). The relationship between the scores on the HDQoL-C and the Perceived Health VAS revealed a Pearsons correlation coefficient of $r(86)=0.055$, $p \geq 0.05$ for component 2 (Practical aspects of Caregiving), $r(86)=0.342$, $p \leq 0.01$ for component 3 (Satisfaction with Life) and $r(86)=0.433$, $p \leq 0.01$ for component 3 (Feelings about Living with HD).

There is therefore, a non-significant and extremely weak positive correlation between the Perceived Health VAS and Component 2 'Practical Aspects of Caregiving', a significant moderate positive correlation between the Perceived Health VAS and Component 3 'Satisfaction with Life' and a significant moderate positive correlation between the Perceived Health VAS and 'Feelings about Living with HD'.

In order to further assess the relationship between perceived health and the HDQoL-C, each of the seven rotated factors (or subscales) subsumed within the above three components were individually correlated with the Perceived Health VAS. This allowed for more in-depth analysis of the relationship between each of the individual subscales and perceived health.

Component 2 of the HDQoL-C comprises three rotated factors (or subscales) relevant to practical aspects of caregiving. The relationship between Subscale 1 (Levels of support and

access to professionals) and the Perceived Health VAS revealed a Pearson's correlation coefficient of $r(86)=0.0.232$, $p \leq 0.01$. Therefore, there is a significant but weak positive correlation between this rotated factor of the component 'practical aspects of caregiving' and the Perceived Health VAS. The relationship between Subscale 2 (Long term and genetic issues) and the Perceived Health VAS revealed a Pearson's correlation coefficient of $r(86)=0.272$, $p \leq 0.01$. Therefore, there is a significant but weak positive correlation between this rotated factor of the component 'practical aspects of caregiving' and the Perceived Health VAS. The relationship between subscale 3 (Daily hassles) and the Perceived Health VAS revealed a Pearson's correlation coefficient of $r(86)=0.009$, $p \geq 0.05$. Therefore, there is a non-significant relationship between this rotated factor of the component 'practical aspects of caregiving' and the Perceived Health VAS.

Component 3 of the HDQoL-C comprises two rotated factors (or subscales) relevant to satisfaction with life. The relationship between Subscale 1 (Overall quality of life issues) and the Perceived Health VAS revealed a Pearson's correlation coefficient of $r(86)=0.344$, $p \leq 0.01$. Therefore, there is a significant weak to moderate positive correlation between this rotated factor of the component 'Satisfaction with life' and the Perceived Health VAS. The relationship between Subscale 2 (Personal issues) and the Perceived Health VAS revealed a Pearson's correlation coefficient of $r(86)=0.239$, $p \leq 0.05$. Therefore, there is a significant weak positive correlation between this rotated factor of the component 'Satisfaction with life' and the Perceived Health VAS.

Component 4 of the HDQoL-C comprises two rotated factors (or subscales) relevant to feelings about living with HD. The relationship between subscale 1 (Negative feelings) and

the Perceived Health VAS revealed a Pearson's correlation coefficient of $r(86)=0.450$, $p \leq 0.01$. Therefore, there is a significant moderate positive correlation between this rotated factor of the component 'Feelings about living with HD' and the Perceived Health VAS. The relationship between Subscale 2 (Positive feelings) and the Perceived Health VAS revealed a Pearson's correlation coefficient of $r(86)=0.265$, $p \leq 0.01$. Therefore, there is a significant weak positive correlation between this rotated factor of the component 'Feelings about living with HD' and the Perceived Health VAS.

The above in-depth analysis of the rotated factors of the HDQoL-C further demonstrates an extremely weak positive correlation between the Perceived Health VAS and Component 2 'Practical Aspects of Caregiving', a significant moderate positive correlation between the Perceived Health VAS and Component 3 'Satisfaction with Life' and a significant moderate positive correlation between the Perceived Health VAS and 'Feelings about Living with HD'. However, by looking at the subscales individually, we are able to establish specific factors that have a non-significant relationship with the Perceived Health VAS. Therefore, those items can be investigated more thoroughly in order to establish their relevance to the scale. For example, although all of the subscales of component 2 demonstrate only a weak relationship with the Perceived Health VAS, factor 3 (Daily hassles) is shown to have a non-significant relationship with perceived health. It would therefore be interesting to further examine the relationship between Daily hassles and perceived health in order to more fully understand the implications of this finding.

Reliability:

The reliability of a scale indicates how free it is from random error. Therefore, carrying out reliability assessments on the HDQoL-C addresses the question of whether the scale can reliably measure the impact of HD on the QoL of family carers. Internal consistency was investigated using Cronbach's Alpha coefficient in order to examine the degree to which the scale items were all measuring the same underlying attribute (i.e. QoL). Test-retest reliability can determine whether the questionnaire is reliable by administering it to a number of participants on two separate occasions and correlating their responses (high-test correlations indicate a more reliable scale as the participant has answered the questionnaire in a similar way on both occasions). To measure the test-retest reliability of the HDQoL-C, 10 carers completed the questionnaire again after a two week period and the correlation between the two scores was calculated.

Internal consistency:

Components 2, 3 and 4 of the HDQOL-C and their subscales were calculated using Cronbach's alpha coefficient.

Table 8.9. below depicts the alpha coefficient of each component and respective subscales.

Table 8.9. Internal Consistency of the HDQoL-C.

	Cronbach's alpha
Component 1: Practical Aspects of Caregiving	0.8010
Subscale 1: Levels of support and access to professionals	0.8009
Subscale 2: Long term and genetic issues	0.8001
Subscale 3: Daily Hassles	0.8007
Component 2: Satisfaction with Life	0.8440
Subscale 2: Overall QoL issues	0.8021
Subscale 2: Personal Issues	0.8140
Component 3: Feelings about Living with HD	0.8850
Subscale 1: Negative feelings	0.8680
Subscale 2: Positive feelings	0.8027

Reliability scores obtained were acceptable as internal consistency should exceed 0.8 (Streiner & Norman, 1995).

Test-retest reliability:

Ten carers completed the HDQoL-C two weeks after first administration. Both questionnaire scores were subsequently correlated using Pearson's correlation coefficient. Table 8.10. below depicts the test-retest reliability of each component and respective subscales.

Table 8.10. Test-retest reliability of the HDQoL-C.

	Pearsons correlation (r=)
Component 1: Practical Aspects of Caregiving	0.78
Subscale 1: Levels of support and access to professionals	0.79
Subscale 2: Long term and genetic issues	0.64
Subscale 3: Daily Hassles	0.81
Component 2: Satisfaction with Life	0.86
Subscale 2: Overall QoL issues	0.84
Subscale 2: Personal Issues	0.87
Component 3: Feelings about Living with HD	0.90
Subscale 1: Negative feelings	0.91
Subscale 2: Positive feelings	0.87

Reliability scores obtained were acceptable as stability should exceed 0.5 (Streiner & Norman, 1995).

Time taken to complete the HDQoL-C and Other Comments.

It took on average 21 minutes to answer the HDQOL-C and carers reported that the questionnaire was user friendly and easy to fill in. This was an important issue as it was considered imperative that the HDQoL-C was a questionnaire that could be filled in quickly and easily in order to be of benefit to the carers and not further restrict their time. Furthermore, a number of the carers reported the cathartic affects of filling in the HDQoL-C. Some commented upon feeling like someone was actually interested in them and what they had to say, which was deemed as very beneficial. Others felt that filling in the HDQoL-

C had given them the opportunity to think about things from their own perspective and allowed them to 'get their feelings out'.

The Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) after Principal components Analysis (PCA) and reliability and validity assessment:

From the 63 items in the pilot version of the HDQoL-C, 27 items were accepted, 4 items were revised, 1 item was rewritten and 31 were discarded. Two qualitative questions were also added to the HDQoL-C in line with comments from experts and carers. There are 34 items in the revised and validated version of the HDQoL-C (see appendix XIII). Table 8.11 outlines the facets and domains of the revised Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) below.

Table 8.11. Domains and facets of the HDQoL-C after PCA and reliability and validity assessment.

DOMAIN (Components)	FACETS (Subscales)
Demographic and Objective Information	N/A
Practical aspects of caregiving	Levels of support and access to professionals Long term and genetic issues Daily Hassles
Satisfaction with life	Overall QoL issues Personal issues
Feelings about living with HD	Negative feelings Positive Feelings

Discussion.

The HDQoL-C has been established as a multidimensional and psychometrically sound disease-specific and subjective QoL assessment tool which incorporates the individual's physical health, psychological state, level of independence, social relationships and personal beliefs. The HDQoL-C demonstrates good internal consistency, test re-test reliability and congruent validity for use with spousal carers of HD patients. It is important to note that the symptoms and genetic nature of HD makes the spousal carer role distinct from other HD family carer roles (e.g Kessler, 1993) and as such, the HDQoL-C may not be a valid and reliable tool for use with other HD-specific carer populations. However, in line with carers' comments with regards to the reported cathartic nature of the questionnaire and experts' comments about its usefulness in terms of giving professionals some 'one to one time' with carers, it is thought that the HDQoL-C may also prove useful in assessing the QoL of other HD family carers, (e.g. child carers, carers who are 'at risk' or carers who are HD positive). As such, it was decided to call the scale the 'Huntington's Disease Quality of Life Battery for Carers' and note that in terms of psychometric properties, further validation is required for its use with other family members. However, even in its current form, it may be of some benefit to such other sub-groups of family carers and health care professionals.

The components on the HDQoL-C show a strong positive correlation with WHOBREF (WHO, 1996). As the WHOBREF is a well validated tool for measuring QoL (WHO group, 1996), the strong positive correlation between the WHOBREF and the components on the HDQoL-C suggests that the HDQoL-C is indeed measuring QoL. As predicted, the correlation although strong, was not perfect and therefore the HDQoL-C may be picking up

on some of the disease specific QoL issues that HD carers face that generic tools do not address.

The components 'Satisfaction with life' and 'Feelings about Living with HD' on the HDQoL-C show a medium positive correlation with the SWLS (Diener, 1985). However, the HDQoL-C component 'Practical aspects of caregiving' only demonstrates a weak positive correlation with the SWLS. This is not surprising as it could be argued that issues surrounding the practical aspects of caregiving in HD will only impact upon the carers QoL if they perceive them as a stressor (Lazarus & Folkman, 1984) or if they do not have the coping mechanisms to deal with such issues.

The components 'Satisfaction with life' and 'Feelings about life' on the HDQoL-C show a medium positive correlation with perceived health status (as measured by VAS, Weinman, 1995). However, the HDQoL-C component 'Practical aspects of caregiving' only demonstrates an extremely weak positive correlation with perceived health status. This further demonstrates that the practical aspects of caregiving may only impact upon the HD-spousal carers health if they do not have the skills to deal with stressors surrounding the practical elements of their care giving roles.

Summary.

In this final study, a HD-specific QoL measure for use with spousal-carers was shown to be both valid and reliable. It is anticipated that in future research the Huntington's Disease

Quality of Life Battery for Carers (HDQoL-C) will be used to gather information regarding the QoL of HD spousal carers and the efficacy of interventions. The scale will also be piloted with other HD specific family caregiving populations in order to expand on its usability.

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1. Research Summary.

This research aimed to systematically investigate the factors that may enhance and compromise the lives of HD spousal carers by utilising the theoretical construct of quality of life (QoL). This investigation provided evidence that spousal carers of HD patients have specific difficulties in maintaining their QoL whilst continuing in a primary care-giving role.

My first study revealed that family carers of HD patients consider the concept of QoL an important issue in HD spousal caregiving. Eighteen sub-themes or issues relating to QoL that were pertinent to the carers emerged from the qualitative data that was collected, which clustered into four final themes of Professional Issues in HD, Personal Wellbeing in HD, Practical Issues in HD and Emotional Wellbeing in HD.

These initial findings provided preliminary evidence that HD does indeed impact on the QoL of family carers. Therefore, it was decided to carry out two more investigative studies that would allow for additional evaluation of the QoL concept in relation to family caregiving in HD. As such, five spousal carers took part in a study that employed the 'photovoice' method in which participants take photographs and make comments on the things in life that are important to them (Aubeeluck & Buchanan, in press). Additionally, the themes that were generated from study 1 and study 2 were taken to six semi-directed focus groups for discussion (Aubeeluck & Buchanan, in preparation).

Analysis of the photovoice data produced nine themes of 'Care and Security', 'Small Pleasures', 'Loneliness', 'Escape', 'Sense of Loss', 'Neglected Needs', 'Support', Lack of

Time' and 'Daily Hassles' that appear to be intrinsically linked with HD spousal carers' QoL. The identified themes lean towards the notion that caring for a family member with HD imposes a unique and difficult burden on the carer's life. Although some positive elements emerged, these appeared to be minimal (i.e. small pleasures like having a cigarette or having flowers in the house) when compared to the negative impact that HD has on their lives. The data suggested that these informal carers often experienced loneliness, a need to escape and a unique sense of loss while trying to adequately care for their loved ones and maintain some form of QoL for themselves.

Analysis of the focus group data supported the findings from studies 1 and 2 with evidence of all of the themes (except for 'small pleasures' identified in study 2), being reported by the carers and health care professionals involved in the discussions. The analysis of the verbatim transcripts identified four additional themes that illustrated the ways in which spousal caregiving in HD impacts on QoL. These were 'Levels of support', 'Dissatisfaction with Caregiving Role', 'Practical Aspects of Caregiving' and 'Feelings and Emotional Wellbeing'. These themes and sub-themes identified in studies 1 and 2, and confirmed in study 3, were able to be integrated into the existing ComQoL-A5 (Cummins, 1997) to generate questions for a pilot version of a HD specific QoL measure for family carers (HDQoL-C).

In the final study, Principal Components Analysis (PCA) of the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) established it as a multidimensional and psychometrically sound disease-specific QoL assessment tool (Aubeeluck, in press; Aubeeluck & Buchanan, in preparation). The HDQoL-C measures the QoL of HD-spousal carers through the subjective interpretation of their physical health, psychological state, level

of independence, social relationships, personal beliefs and relationship to salient features of the environment. The HDQoL-C demonstrates good internal consistency, test re-test reliability and congruent validity.

2. General Conclusions

This current research and the extant Dementia Caregiving Literature.

Within the dementia caregiving literature it has been firmly established that family members play a leading role in homecare of the demented patient (Shanas, 1979; Cantor, 1983; Johnson, 1983). A number of dementia caregiver studies demonstrate the personal, health and social impacts of dementia care and the financial burden that it places upon the family (e.g. Almberg, Grafstrom & Winblad, 1997; Clark & Bond, 2000; O'Connor et al, 1990). The studies in this thesis provide evidence that family carers of people with HD also experience an impact of caregiving on their overall wellbeing. HD carers report feelings of burden and exhaustion and increased levels of stress. Furthermore, they describe financial concerns, family conflict and time constraints. These issues also appear to be impacted upon by the genetic nature of HD. Caregivers may be individuals who are 'at-risk' or HD positive themselves. Or, they may have children who are 'at-risk' who will require care in years to come. Moreover, the genetic status of family members may also lead to wider financial concerns as individuals who have tested positive for HD are obliged to divulge such information to insurance companies. As such, families can find themselves in a situation where they have no life insurance, and loan and mortgage applications are refused. Such

issues mean that HD permeates the entire life of the family carer as they cope with a chronic caregiving situation that is impacted upon through financial worries and concerns.

The role of the HD family carer appears to be comparable to that of the dementia family carer in terms of the personal and social burden that it places upon carers and the health implications it may have. However, caregiving in HD appears to bring with it the unique burden of dominant genetic inheritance. HD is a family disease in which carers may find themselves looking after a number of family members at one time or caring over a number of generations with very little financial support. As such, stress-process models of caregiving in dementia may prove useful in furthering our understanding of caregiving in HD.

This current research and the extant HD Caregiving Literature.

As with the general dementia caregiving literature, it is generally the immediate family that takes on the responsibility of caring for an HD affected individual. In terms of the symptomology of HD, there are many issues and concerns that were highlighted in this thesis that fall in line with the existing literature on caregiving in HD. For example, Power (1982) notes how cognitive dysfunction in HD can lead to patients becoming apathetic and inactive preferring to say at home, and placing a huge burden on the carer. The carers that took part in focus group sessions, also reported their frustration with their HD affected family members with many commenting upon how difficult it was for them to motivate their relative and the increased stress that they experienced because of this. In the same study, Power (1982) also noted some discomfort and embarrassment from

family members in relation to going out with their HD affected relative, and the response that the movement disorder would generate from the general public. However, within this sample, carers reported situations in which they had experienced feelings of empathy and acceptance by members of the general public. Carers further stated that behaviour problems such as apathy were often more difficult for them to cope with than the movement disorder and any embarrassment that came with it. This suggests that the carers in this thesis are more concerned with the impact that HD is having on their relationships, as changes in behaviour impact on the dynamics of the family and can cause feelings of tension and stress. It should also be considered that over the last 20 years since Power's study, general attitudes towards disease and illness have changed and that the stigma that was once associated with diseases such as HD may not be as prevalent now as they were then.

Hayden et al (1980) demonstrated the burden that HD places on the family. Within their sample, carers talked about the burden of carergiving and the associated guilt that they experienced from feeling burdened. Carers felt that they should be able to cope with the situation they had been placed in and saw their feelings of burden as a sign that they were not coping as well as they should be, which in turn led to feelings of guilt. Indeed, guilt was an emotion that was continually referred to by the carers in this study, they felt guilty when they couldn't cope, when they went out and did something for themselves and even when they talked about the difficulties they were experiencing with other people.

Dura (1993) found that the long-term effects of educational interventions for carers of

HD patients were minimal due to the stressfulness of continuing to provide care in this insidious and chronic disorder. However, the carers in this sample were keen to find out more about HD and ways to cope and deal with situations. Many carers saw education as the 'key' to some of their problems and believed that it was highly beneficial to them. This changing opinion with regards to the usefulness of education, may be in some part due to the current environment in which many carers have access to a vast array of educational resources thorough the internet. Indeed, recent research in relation to HD patients and carers and their use of the Internet (Coulson, Buchanan, Aubeeluck & Rooney, in submission) has established that HD patients and carers do see knowledge as a key element of coping and use the internet as a source of education and support.

Hans & Gilmore (1968) note the major emotional, social and financial problems that caregiving in HD creates for the family and that such issues are made worse due to lack of attention that HD has received from public health services in terms of interventions. The carers in these current studies also raised concerns with regards to the emotional burden they experienced, the lack of time they had for themselves which impacted upon their social life and the huge financial problems that having a genetically inherited disease imposes on the family. Moreover, patients and their families found tremendous difficulty in gaining access to specific services and felt that the professionals they dealt with did not always have an in-depth knowledge of HD.

The evidence from this thesis therefore supports previous findings in relation to caregiving in HD. It would appear that HD spousal carers play a vital role in supporting their HD affected family members. However, caregiving is demanding, and levels of

stress and self-reported burden and distress among carers are high. The needs of HD carers therefore need to be addressed, high levels of stress and burden require identification and appropriate interventions or support require implementation. Where this is not the case, health services may find themselves with two patients to support where previouslythere was only one, supported by the HD carer.

This current research within the context of extant QoL research.

Existing research has seen QoL emerge as a useful outcome measure by which to judge the efficacy of psychological interventions (e.g. Cummins, 1997; Land, 2000; Rapley, 2003). However, there are many documented concerns about the conceptualisation of QoL (e.g. Cummins et al, 1994; Allison, Locker & Feine, 1997; Anderson & Burckhardt, 1999) and as such, about the usefulness of the concept itself. Nonetheless, authors such as Cummins (1997) argue that these concerns can often be addressed through the use of a well operationalised and validated tool.

This current research has attempted to overcome some of these barriers and concerns in relation to the measurement of QoL. By operationalising and re-defining QoL throughout the research, all the data that has been generated within the studies should fall in line with current prevalent definitions of QoL. Moreover, by taking a 'bottom up' approach to the measurement of QoL and seeing the individual as the 'expert' within their own situation, the HDQoL-C has been developed by the user for the user, rather than through a 'top down' academic approach. Using the HDQoL-C as a measure of outcome has the advantage of focusing on the impact of a HD on the *individual's*

emotional and physical functioning and lifestyle. Therefore, as a tool that has been developed in conjunction with the individuals who are ultimately to use it, the HDQoL-C should provide a reliable and client-led baseline against which the effects of any intervention can be evaluated.

It is however important to note that there is no agreed definition or standard form of measurement of QoL and as such, the QoL construct can become very complex in composition. Moreover, there is little empirical research attempting to define those qualities which make life and survival valuable. The literature covers a range of components from which QoL is often derived, such as functional ability, including role functioning (e.g. domestic, return to work), the degree and quality of social and community interaction, psychological well-being, somatic sensation (e.g. pain) and life satisfaction. The findings from this current research provide further evidence of the difficulties in defining the components and qualities that are relevant to the QoL of the HD spousal caregiver. In the item analysis stage of the development of the HDQoL-C, it became apparent through a series of correlation's between the sub-components of the HDQoL-C and an existing reliable and valid QoL scale, that some elements of the HDQoL-C correlated more strongly with the existing scale than others. This may reflect the fact the HDQoL-C has been developed as a disease-specific tool and in order to validate it, it has been correlated with an existing generic scale. As such, it would be expected that some disease-specific sub-components would correlate with the generic scale more strongly than others. Conversely, it could be argued that differences within inter-factor correlation's are due to the nature of QoL and the difficulties in establishing the specific issues that are pertinent to its measurement.

Quality of life is a useful construct to use in outcome intervention research. However, there are still many methodological and conceptual issues that are being debated with regards to the operationalisation of QoL, its objective and subjective components and further, the use of disease and population specific scales vs more generic determinants of QoL. However, it is generally accepted that there is a need for both objective measures of QoL that assess particular tangible needs and subjective indicators, in order to allow the clinician or researcher to gain understanding of the impact of a situation on the *individual.* This current research has attempted to combine the objective with the subjective in order to provide a holistic account of HD spousal caregivers situation.

Finally, the use of the HDQoL-C as a disease -specific measures of QoL can be a useful and beneficial supplement to more generic QoL measures that may not be sensitive to QoL issues that surround caregiving in HD. By combining disease-specific and generic QoL assessment tools, HD-related issues can be assessed whilst findings can still be generalised to other populations.

Methodological Issues

Using Factor Analysis with Small Sample Sizes.

An issue that is frequently debated when using factor analysis is the sample size required for obtaining a reliable result. Small sample sizes may negatively affect the outcome of the factor analysis procedure and a number of researchers have given guidelines for the minimum sample size needed to conduct factor analysis. Some have suggested the ratio of sample size

to number of variables as a criteria with recommendations ranging from 2:1 through to 20:1 (e.g. Baggaley, 1982, Marascuilo & Leven, 1983). Others have suggested a minimum sample size of 100 to 200 participants (e.g. Linderman et al, 1980; Guadagnoli and Velicer, 1988). The sample size in this study was 87, just below the recommended minimum of 100 indicating that the small sample size may be a problem. However, the concern with small sample sizes is that they may affect the factor analysis by making the solution unstable i.e. the addition of more data may cause the variables to switch from one factor to another (Guadagnoli and Velicer, 1988). However, due to the low prevalence rates of HD, the data collected for this study is from a large proportion of the UK HD spousal carer population. As such, sample size effects are not likely to be present within the data.

Measuring the QoL of Spousal Carers of HD patients.

There are many documented concerns about the construct of QoL and how it should be measured (Rapley, 2003). The main concerns that arise tend to be with regards to the use of generic quality of life measures with disease-specific populations. Moreover, issues in terms of whether to measure QoL objectively or subjectively make problematic the value of QoL measures and their generalisability and application, both within and between different populations. However, there is evidence to suggest that specific caregiver populations may benefit from specially adapted QoL measures (Coon, 2002), in order that disease and population-specific issues are 'tapped' into.

The multi-faceted nature of HD and evidence from my studies do suggest that HD spousal carers struggle to maintain an acceptable level of life quality due to the full time care they provide. The objective of developing the Huntington's Disease Quality of Life Battery for

Carers (HDQoL-C) was therefore, to quantify the caregiving experience and set the most beneficial and cost-effective support in place via a self-report questionnaire. It should be noted that the symptoms and genetic nature of HD make the spousal carer role distinct from other HD family carer roles (e.g Kessler, 1993) and as such, the HDQoL-C may not be a valid and reliable tool for use with other HD-specific carer populations. However, the HDQoL-C showed excellent face validity with a variety of family carers commenting upon its relevance to their own experiences. Therefore, it is thought that the HDQoL-C may also prove useful in assessing the QoL of other HD family carers, (e.g. child carers, carers who are at risk or carers who are HD positive). However, further validation is required for its use with these specific populations.

Using the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) as an outcome measure in research.

Bowling (2001) notes that purchasers of health care are generally expected to allocate resources on the basis that such resources are indeed advantageous to the individual and also, effective. As such, QoL interventions need to have some proved 'health gain' in order for them to be seen as effective and beneficial. Using the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) as a measure of outcome has this benefit of focussing on the impact of HD on the *carers'* emotional and physical functioning and lifestyle. As such, used as an outcome measure, the HDQoL-C may help to answer the question of whether an intervention leads to an increase in wellbeing by providing a more individual-led baseline against which the effects of the intervention can be evaluated.

It is anticipated that as the HDQoL-C has been developed through both theory and data driven methods, and has been shown to be psychometrically sound, it will prove effective in the assessment of HD spousal carer interventions. As such, it may help to build upon our current understanding of the issues surrounding caregiving in HD and establish effective ways of alleviating HD caregivers' distress, therefore helping to improve QoL. Furthermore, it is hoped that as studies are carried out and interventions are found to be effective (or ineffective), the psychological literature with regards to the impact of HD on the QoL of spousal and family caregivers will expand.

Using Mixed Methodology in Quality of Life Research.

This current study has utilised a number of established approaches as well as more novel methods in the development of a scale to measure the impact of HD on the QoL of spousal carers. In the initial stages of scale development it was decided to take an existing generic QoL measure and assess it for its relevance to the QoL of the spousal caregiver. There are already a number of existing tools that measure caregiver distress in dementia, however the purpose of this thesis was develop a scale that took a holistic approach to the measure of life quality, encompassing both positive and negative dimensions rather than just tapping into the construct of caregiver distress. As such, QoL and its theoretical underpinnings became the foundation of the research rather than the existing literature on caregiver distress in dementia. It should therefore be acknowledged that had the scale development begun from

a caregiving background, the final scale and its sub-components may have been slightly different with more of a focus on caregiver distress rather than QoL.

The second study in this thesis used a novel approach known as 'photovoice' in order to generate subjective information about the impact of HD on the QoL of the spousal carer. 'Photovoice' provided a unique opportunity for HD carers to capture and reflect on issues relating to QoL as they arose on a daily basis, which may not have been possible using other methods. It also allowed carers the opportunity to reflect on their caring role but required minimal time out of their daily routines. Moreover, taking part in 'photovoice' appeared to be of therapeutic benefit to the carers who took part in the study. Carers wrote notes of thanks at the end of the validation study reporting how much they had benefited from filling out the HDQoL-C. They commented upon how it had allowed them to think about their situation from their own perspective rather than thinking about the patient all of the time. They also noted that the mere existence of the questionnaire had made them feel that someone cared about them, and furthermore, that someone might listen to them and be able to help them. However, it is important to note that only a small and self-selecting sample of carers took part in this study and this may well have influenced the findings. Moreover, content analysis of the data may have been subject to experimenter bias as previous knowledge may have influenced the way the data was categorised.

Study three aimed to overcome some of these issues by taking the themes generated in the first two stages of research to a larger group of HD carers and discussing them within a focus group setting. This allowed for issues to be further debated by HD carers before the scale items were constructed, thus, taking a 'bottom up' approach to the research and

acknowledging the carer as the 'expert' of their caregiving situation. This was a good way of clarifying the importance of a number of issues and their impact on QoL and was incredibly useful in the construction of the scale and wording of items. However, it is again important to note that the carers who took part in the focus groups may not be representative of the HD carer population as a whole. For example, it may have been difficult to tap into some of the issues and concerns of the most burdened caregivers as this particular sub-group may find it difficult to find the time to nd take part in a focus group session.

The final study in this thesis was the validation of the scale items that had been generated within the previous qualitative research. The questionnaire was piloted with 87 HD spousal carers and principal components analysis was performed on the data in order to generate a concise and accurate scale that can measured the QoL of the HD spousal carer. By combining qualitative methodology in the generation of the scale items and refining them through statistical analysis, the HDQoL-C has been developed by HD carers as experts of their own situation and confirmed through well established statistical methods in order to produce a user friendly tool that taps into issues surrounding the impact of HD on the QoL of spousal carers.

Future directions.

Validation of the HDQoL-C in other HD populations.

A version of the Huntington's Disease Quality of Life Battery (HDQoL-C) has been developed for use with family carers of Juvenile HD (JHD) patients in conjunction with the

Huntington's Disease Association (HDA) UK. JHD is a very rare condition with only about 10 percent of HD cases occuring in individuals under the age of 20 years (Rasmussen et al, 2000). JHD or early-onset HD has an onset age of anywhere between infancy and 20 years, with the youngest patient described in the literature, having an onset age of 2 years (Huntington's Disease Collaborative Research Group, 1993). Although JHD and adult-onset HD both result from an altered form of the huntingtin gene, the symptoms of JHD are very different from those of adult-onset HD. Individuals with JHD often become stiff or rigid in their movements (instead of having chorea) and about one third experience recurrent seizures (Hayden, 1981). As with adult-onset HD, individual cases of JHD vary greatly, and different children often have different symptoms. The earlier the onset of JHD, the faster it usually progresses and in general, progression of the disease is more rapid than in adult-onset HD. Often, death from JHD occurs within 10 years of onset, as opposed to 10-25 years in adult-onset HD with JHD being a more severe disease than in adult cases (Kremer, 2002).

Because of its hereditary nature of JHD and early age of onset, a child with JHD may also have a parent or other close family member who is affected by adult-onset HD at the same time. This tendency to affect multiple generations simultaneously places an even greater strain upon families who are affected by juvenile HD. As such, caring for a child with JHD brings with it additional burdens in relation to possible dual caregiving roles and further the emotional bereavement and loss often experienced by parents when caring for a terminally ill child (Gravelle, 1997). In order to make the HDQoL-C relevant to this specific HD carer population, a number of changes were made. Table 9.1 outlines these changes.

Table 9.1. Changes made to the HDQoL-C for use with carers of Juvenile Huntington's Disease Patients.

	Original Version	JHD carer version
Q 1d	" What is your marital status?"	Additional Item added: "widowed"
Q1e	"How long have you known of the presence of HD in your family?"	"IS HD in your family or in your partners family?
Q1f	"How long have you being caring for an HD affected family member?"	"How long have you being caring for an JHD affected family member?"
Q1h	"What relation to you is the HD affected family member you are caring for?"	"What relation to you is/ are the HD affected family member(s) you are caring for?"
Q1j	"Do you have any children?"	"How many children do you have and what is their genetic status" followed by boxes to put number in - How many are at risk? How many have tested positive? How many are not at risk?"
Part 2 Q5	"How often do the genetic implications of HD impact upon your caregiving role"	" How often can you see no end to your caregivng role due to the genetic nature of HD?"
Part 4Q9	"I feel worried about the genetic implications of HD.	"I feel worried that our family will never be free from HD because of its genetic nature"

The Juvenile Huntington's Disease Quality of Life Questionnaire for Carers (JHDQoL-C) (See appendix XI) has been distributed to 30 Carers of patients with JHD. It is hoped that once validated, the JHDQoL-C will be used widely within HD clinics across the UK.

Validation of the HDQoL-C in other cultures.

A North American version of the Huntington's Disease Quality of Life Battery (HDQoL-C) has been developed and is currently being piloted in conjunction with the University of Columbia in North America. In order to make the HDQoL-C user friendly within the North American population, some small language amendments were made. Table 9.2 outlines the changes that were implemented.

Table 9.2 Changes made to the HDQoL-C for use with a North American Population.

	UK version	US version
Title	The term 'Battery' is used	changed to 'Questionnaire' as the term 'Battery' is not used in the US
Example question	"... .support that you need from others?"	changed to "... support that you need"
Explanation added		Text added: The HD affected family member is sometimes referred to as a "person with HD" or simply pHD.
Q1c	"What is the highest education you received?"	"How many years of education do you have?
Q1e	"How long have you known about the presence of HD in your family?	"How long have you known about HD in your family?"
Q1j	"Do you have any children? If so, what is their genetic status?"	"Do you have any children? If YES, how many? (followed by boxes to put number in - How many are at risk? How many have chosen to be tested for the HD gene? How many have chosen to not be tested?" >
Q1k		Addition item: "How many family members live in your household? Followed by boxes to put numbers in.
Q 3		Additional item: list of common medical conditions e.g. hypertension, high cholesterol, depression, diabetes mellitus, cardiac, bone or muscle pain.
Q4	"How many hours do you spend on the following each week?"	"How many hours do you spend on the following activities each week?"
Q4	"Hours paid work"	"Hours paid to work outside the home"
Q4		Additional Item: "Hours paid to work caring for pHD"
Q4		Please specify any ***difficulties you experience*** caring for your pHD,e.g. Coping with behavioral problems such as: Irritability and temper outbursts, Lack of initiation, Communication difficulties, Coping with physical problems such as: Chorea, Rigidity, Swallowing difficulties. Coping with emotional problems such as Depression, Mania, Sexual disorder.
Q5	"... Utility Room"	"... .Laundry Room"

The Huntington's Disease Quality of Life Questionnaire for Carers, North American Edition (HDQoL-C.US) (See appendix XII) has been distributed to 60 North American Carers of patients with HD and at present 17 completed questionnaires have been returned. It is hoped that once validated, this version of the HDQoL-C will be used widely within HD clinics across the US.

Using the HDQoL-C as an outcome measure for Therapeutic Interventions.

The purpose of assessing the HD spousal carers situation is to gain an understanding of their role that may help to prevent or to ameliorate QoL-related problems, enabling the carer to maintain a good standard of life quality whilst continuing to provide care comfortably for their relative. The basis of successful intervention therefore, lies in gaining an informed understanding of the individual case. As such, using the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) may provide helpful ways of organising the information that is gained during assessment. For example, the HDQoL-C will provide the health care professional with the carer's objective QoL information such as financial situation or, the practical suitability of the home as well as subjective QoL information such as feelings of stress or depression. As such, the HDQoL-C will demonstrate possible areas for intervention at the level of increasing both objective and subjective QoL in spousal carers of HD patients.

Furthermore, anecdotal evidence that has been gathered during the course of these studies has suggested that just completing the HDQOL-C may have therapeutic and cathartic benefits for HD carers. The majority of carers wrote notes of thanks at the end of the pilot

questionnaire saying how much they had benefited from filling out the HDQoL-C. They commented upon how it had allowed them to think about their situation from their own perspective rather than thinking about the patient all of the time. They also noted that the mere existence of the questionnaire had made them feel that someone cared about them, and furthermore, that someone might listen to them and be able to help them. From the clinician's or health care professionals' viewpoint, even using the HDQoL-C at this level may be very beneficial to his / her relationship with the HD carer.

Continuing Research into Family Caregiving in HD within the field of Health Psychology.

The issues that have emerged from these current studies raise a number of concerns with regards to the importance of recognising the needs of HD spousal caregivers. Therefore, it is paramount that service providers, researchers and policy makers identify the best way to assist carers by taking into account their differing needs in relation to stressors that spousal caregiving in HD seem to impose on the individual. With the general remit of health psychology being to promote well-being via the application of psychological models and theories, the issue of caregiving in HD is undoubtedly an area in which the health psychologist can have an impact. For example, through the practical application of designing self-care programs, using therapeutic interventions or by advocating carers' views on service quality.

Kessler (1993) notes that there is a paucity of research investigating the overall QoL of carers in the HD literature. In addition, non-HD specialist professionals such as general nurses, GP's or social workers may be unaware of the tremendous impact that HD has on family life. The complex nature of HD and its subsequent repercussions on the family make it unlikely that any one professional will have all the skills needed to help an individual family. As such, certain elements of care are bound to be passed on to staff who may have little or no previous experience of working with HD patients and their families (Shakespeare and Anderson, 1993). It is hoped therefore, that the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) will provide a an understanding of the factors that impact upon the HD spousal carers' QoL in order to aid health care providers in their assessment and any subsequent intervention that may be necessary to improve the HD spousal caregiver's situation.

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Appendix III. The Huntington's Disease Quality of Life Questionnaire for Carers (HDQoL-C) - Pilot Version.

Huntington's Disease Quality of Life Scale for Carers.

(HDQoL-C)

Aimee Aubeeluck and Heather Buchanan

School of Education, Health and Sciences
University of Derby

Correspondence to:

Aimee Aubeeluck
Centre for Psychological Research in Health and Cognition
School of Education, Health Care and Sciences
University of Derby
Western Road
Mickleover DE3 9GX
Tel: 0870 040 7385

e-mail: a.aubeeluck@derby.ac.uk

HDQoL-C

Thank you for taking the time to fill in this questionnaire. The questionnaire has three sections. The first will ask for some factual information. The next two will ask how satisfied you are and how you feel about various aspects of your life.

We want to know how you feel about your quality of life, your health and other areas of your life. **Please answer all the questions.** If you are unsure about which response to give to a question, **please choose the ONE** that seems most appropriate (this is often your initial response).

Where there is an 11 point scale (0-10), please circle the number that you feel most accurately represents your situation. For example, a question might ask:

Q: Do you get the kind of support from others that you need?

Almost never Almost always

0 1 2 3 4 5 6 7 8 9 10

You should **circle** the number that best fits the kind of support you get. So if you very rarely get the kind of support from others that you need, you would circle the number 1.

Section 1.

This section asks for information about various aspects of your life. Please answer all the questions and do not spend too much time on any one item.

1a) What is your date of birth? ... ...

b) What is your sex ? (please circle) Male Female

c) What is the highest education you received ?
Primary school
Secondary school
University
Post-graduate

d) What is your marital status?
Single
Married
Living as married
Separated
Divorced
Widowed

e) How long have you known of the presence of HD in your family? ... ...

f) How long have you been caring for an HD affected family member? ... ...

g) Are you the main carer for your HD affected family member? YES NO

h) What relation to you is the HD affected family member you are caring for? (e.g. spouse, sister, parent etc) ... ...

i) Have you previously cared for any other HD affected family members? YES NO

- *if so*, what was their relationship to you? (e.g. spouse, sister, parent etc) ... ...

2. **What is your personal or household (whichever is most relevant to you) gross annual income before tax?**

£_____

3. **How many times have you seen a doctor over the past 3 months?**

None	1 – 2	3-4 (about once a month)	5-7 (about every two weeks)	8 or more (about once a week or more)
☐	☐	☐	☐	☐

4. **Do you have any disabilities or medical conditions? (e.g. visual, hearing, physical, health, etc.).**

Yes ☐ No ☐

If yes please specify:

Name of disability or medical condition	*Extent of disability or medical condition*
e.g. Visual	Require glasses for reading
Diabetes	Require daily injections
Epilepsy	Requires daily medication
_____	_____
_____	_____
_____	_____

5. **How many hours do you spend on the following *each week*? (Average over past 3 months)**

	0	1-10	11-20	21-30	31-40+
Hours paid work	☐	☐	☐	☐	☐
Hours unpaid caring	☐	☐	☐	☐	☐
Hours unpaid child care	☐	☐	☐	☐	☐

6. **In your opinion, is your home suitable / suitably adapted for your family's needs?**

Not suitable									Very suitable	
0	1	2	3	4	5	6	7	8	9	10

7. **Below is a list of leisure activities. Indicate how often in an *average month* you attend or do each one for your enjoyment (not employment).**

Activity		Number of times you do activity each month
(1)	Go to a club/group/society	_____
(2)	Go to a hotel/bar/pub	_____
(3)	Watch live sporting events (Not on TV)	_____
(4)	Go to a place of worship	_____
(5)	Chat with neighbours	_____
(6)	Eat out	_____
(7)	Go to the cinema	_____
(8)	Visit family or friend	_____
(9)	Play sport or go to a gym	_____
(10)	Other activities	_____

- Please tell us what these other activities are:_____

8. **A) Do you have difficulty coping with caring for someone with HD?**

Almost always									Almost never	
0	1	2	3	4	5	6	7	8	9	10

B) Please list any difficulties that you have (e.g. dealing with behaviour, physical problems, emotional problems etc)

_____

_____

_____

_____

9. How EASY is it to get access to any information that you may need about HD or caregiving?

Very easy — incredibly difficult

0 1 2 3 4 5 6 7 8 9 10

10. How important is it for you to maintain a regimented daily routine?

Very important — unimportant

0 1 2 3 4 5 6 7 8 9 10

11. How often do you receive appropriate help from social services?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

12. How often do you have access to professionals who have specialised knowledge of hd and understand its implications?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

13. How often do you have access to appropriate specialist services?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

14. How often do you receive support for yourself when you need it?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

15. How much support are you given by health care professionals?

As much a I need — None Whatsoever

0 1 2 3 4 5 6 7 8 9 10

16. How often do people fight your rights as a caregiver?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

17. How often do the genetic implications of HD impact upon your caregiving role?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

18. How often do you have access to appropriate care facilities?

Whenever you need them — Almost never

0 1 2 3 4 5 6 7 8 9 10

19. How often do you receive any practical support that you need?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

20. How often do you experience a conflict of interest between what you want and what your HD affected relative wants?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

21. How often do you have some spare time?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

22. How often do you talk with a close friend?

Daily — Less than once a month

0 1 2 3 4 5 6 7 8 9 10

23. If you are feeling sad or depressed, how often does someone show they care for you?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9
10

24. If you want to do something special, how often does someone else want to do it with you?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9
10

25. How often do you sleep well?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9
10

26. Are you safe and secure at home?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9
10

27. How often are you worried or anxious during the day?

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

28. How often can you do the things you *really* want to do?

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

29. When you wake up in the morning, how often do you wish you could stay in bed *all day?*

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

30. How often do you have wishes that *cannot* come true?

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

Section 2

This section asks how *satisfied* are you with each of the following life areas.

There are no right or wrong answers. Please circle the number that best describes how **satisfied** you are with each area of your life.

1. How *satisfied are you* with your HEALTH?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

2. How *satisfied are you* with what you ACHIEVE IN LIFE?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

3. How *satisfied are you with* your CLOSE RELATIONSHIPS WITH FAMILY OR FRIENDS?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

4. How *satisfied are you* with HOW SAFE YOU FEEL?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

5. How *satisfied are you* with FEELING A PART OF YOUR COMMUNITY?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

6. How *satisfied are you* with YOUR OWN HAPPINESS?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

7. How *satisfied are you* with THE TREATMENT THAT YOUR HD affected relative RECEIVES?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

8. How *satisfied are you* with YOUR OVERALL QUALITY OF LIFE?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

Section 3

This section asks how you *feel* about each of the following life areas.

There are no right or wrong answers. Please circle the number that best describes how you feel about each area of your life.

1. I *feel* BETRAYED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

2. I *feel* LONELINESS

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

3. I *feel a* SENSE OF LOSS

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

4. I *feel* GUILTY

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

5. I *feel* that the FUTURE IS BLEAK

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

6. I *feel* FINANCIALLY DISADVANTAGED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

7. I *feel* DEPRIVED OF A HELPMATE / PARTNER

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

8. I *feel* ISOLATED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

9. I *feel* STRONG

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

10. I *feel* HOPE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

11. I *feel* SUPPORTED BY HEALTH CARE PROFESSIONALS

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

12. I *feel* FRUSTRATED

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

13 I *feel* a BURDEN of RESPONSIBILITY

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

14. I *feel* that I have LOST MY IDENTITY

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

15. I *feel* EXHAUSTED

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

16. I *feel* SUPPORTED BY FAMILY AND FRIENDS

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

17. I *feel* SAD OR DEPRESSED

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

18. I feel ANGRY

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

19. I feel STRESSED

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

20. I feel EMOTIONALLY DRAINED

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

21. I *feel* WORRIED ABOUT THE GENETIC IMPLICATIONS OF HD

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

22. I *feel* THE NEED TO ESCAPE MY CAREGIVING ROLE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

23. I *feel* LIKE THERE ARE NOT ENOUGH HOURS IN THE DAY.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

24. I *feel* LIKE MY OWN NEEDS ARE NOT IMPORTANT TO OTHERS.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

25. I *feel* COMFORTED BY THE BELIEF THAT ONE DAY THERE WILL BE A CURE FOR HD

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

26. In some ways I *feel* THAT HD HAS HAD A POSITIVE IMPACT ON MY LIFE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

27. I *feel* COMFORTED BY RELIGIOUS BELIEFS

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

28. I *feel* THAT I CAN COPE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

29. I *feel* THAT HD HAS MADE ME A STRONGER PERSON.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

30. I *feel* THAT I HAVE RESIGNED MYSELF TO A LIFE OF CARING.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

31. I *feel* FRUSRTATED BY THE DAILY HASSLES OF CARING.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

32. I *feel* THAT THERE ARE TOO MANY SECRETS IN MY FAMILY.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

33. I *feel* OVERWHELMED BY MY CAREGIVING ROLE.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

34. I *feel* THAT I HAVE HAD A 'DUTY OF CARE' FORCED ON TO ME.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

35. I *feel* LIKE I DON'T KNOW WHO I AM ANYMORE.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

Thank you for your time.

Appendix VIII. The Juvenile Huntington's Disease Quality of Life Questionnaire for Carers (JHDQoL-C) - Pilot Version.

The Juvenile Huntington's Disease Quality of Life Battery for Carers (JHDQoL-C)

Aimee Aubeeluck and Heather Buchanan

School of Education, Health and Sciences

Correspondence to:

Aimee Aubeeluck
Centre for Psychological Research in Health and Cognition
School of Education, Health Care and Sciences
University of Derby
Western Road
Mickleover DE3 9GX
Tel: 0870 040 7385
e-mail: a.aubeeluck@derby.ac.uk

Thank you for taking the time to fill in this questionnaire. The questionnaire has four sections. The first section will ask for some factual information. The next three will ask about different aspects of your role as a carer, how satisfied you are and how you feel about various aspects of your life.

We want to know how you feel about your quality of life, your health and other areas of your life. **Please answer all the questions.** If you are unsure about which response to give to a question, **please choose the ONE** that seems most appropriate (this is often your initial response).

Where there is an 11 point scale (0-10), please circle the number that you feel most accurately represents your situation. For example, a question might ask:

Q: Do you get the kind of support from others that you need?

Almost never								Almost always		
0	1	2	3	4	5	6	7	8	9	10

You should **circle** the number that best fits the kind of support you get. So if you very rarely get the kind of support from others that you need, you would circle the number 1.

Section 1.

This section asks for information about yourself. Please answer all the questions and do not spend too much time on any one item.

1a) What is your date of birth? ……..

b) What is your sex ? (please circle) Male Female

c) What is the highest education you received?
Primary school
Secondary school
University
Post-graduate

d) What is your marital status?
Single
Married
Living as married
Separated
Divorced
widowed

e) Is HD in your family or in your Partners family? ……..

f) How long have you been caring for an JHD affected family member? ……..

g) Are you the main carer for your HD affected family member? YES NO

h) What relation to you are the HD affected family member(s) you are caring for? ……..

i) Have you previously cared for any other HD affected family members? YES NO

- *if so*, what was their relationship to you? (e.g. spouse, sister, parent etc) ……..

j) How many children do you have? ……..

- *AND* what is their genetic status?

How many are at risk?	How many have tested positive for the HD gene?	How many are not at risk?

2. What is your personal or household (whichever is most relevant to you) gross annual income before tax?

£_____

3. Do you have any disabilities or medical conditions? (e.g. visual, hearing, physical, health, etc.).

Yes ☐ No ☐

If yes please specify:

Name of disability or medical condition	*Extent of disability or medical condition*
e.g. Visual	e.g. Require glasses for reading
Diabetes	Require daily injections
Epilepsy	Requires daily medication

_____

_____

4. How many hours do you spend on the following *each week*? (Average over past 3 months)

Hours paid work	0 ☐	1-10 ☐	11-20 ☐	21-30 ☐	31-40+ ☐
☐urs ☐d child care	0	1-10	11-20 ☐	21-30 ☐	31-40+ ☐
☐urs ☐ caring for pHD	0	1-10	11-20 ☐	21-30	31-40+ ☐

Please specify any ***difficulties you experience*** caring for your person with Huntington's Disease (pHD) (e.g. dealing with behaviour, physical problems, emotional problems etc)

_____

_____

_____

5. Is your home suitable / suitably adapted for your family's needs?

Yes ☐ No ☐

If No please specify:

Area of concern	*Problem that it causes*
e.g. No stair lift	e.g. Difficulty moving patient
No utility room	Difficulty getting the laundry done

_____

_____

6. Below is a list of leisure activities. Indicate how often in an *average month* you attend, or take part in, each one for your enjoyment (not employment).

Activity		Number of times you do activity each month
(1)	Go to a club/group/society	_____
(2)	Go to a hotel/bar/pub	_____
(3)	Watch live sporting events (Not on TV)	_____
(4)	Go to a place of worship	_____
(5)	Chat with neighbours	_____
(6)	Eat out	_____
(7)	Go to the cinema	_____
(8)	Visit family or friend	_____
(9)	Play sport or go to a gym	_____
(10)	Other activities	_____

- Please tell us what these other activities are:

_____

Section 2.

This section asks for information about different aspects of your role as a carer.
Please circle the number that you feel most accurately represents your situation. For example, a question might ask:

Q: Do you get the kind of support from others that you need?

Almost never — Almost always

0 1 2 3 4 5 6 7 8 9 10

You should **circle** the number that best fits the kind of support you get. So if you always get the kind of support from others that you need, you would circle the number 10.

---

1. How often are you restricted by the need to maintain a regimented daily routine?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

2. How often do you receive appropriate help from social services?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

3. How often do you have access to professionals who have specialised knowledge of HD and understand its implications?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

4. How much support are you given by health care professionals?

As much a I need — None Whatsoever

0 1 2 3 4 5 6 7 8 9 10

5. How often can you see no end to your caregiving role due to the genetic nature of HD?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

6. How often do you have access to appropriate care facilities?

Whenever you need them — Almost never

0 1 2 3 4 5 6 7 8 9 10

7. How often do you receive any practical support that you need?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

8. How often do you experience a conflict of interest between what you want and what your HD affected relative wants?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

9. How often do you sleep well?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

Section 3.

This section asks how *satisfied* are you with each of the following life areas.

Please circle the number that best describes how **satisfied** you are with each area of your life.

---

1. How *satisfied are you* with your HEALTH?

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

2. How *satisfied are you* with what you ACHIEVE IN LIFE?

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

3. How *satisfied are you with* your CLOSE RELATIONSHIPS WITH FAMILY OR FRIENDS?

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

4. How *satisfied are you* with HOW SAFE YOU FEEL?

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

5. How *satisfied are you* with FEELING A PART OF YOUR COMMUNITY?

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

6. How *satisfied are you* with YOUR OWN HAPPINESS?

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

7. How *satisfied are you* with THE TREATMENT THAT YOUR HD affected relative RECEIVES?

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

8. How *satisfied are you* with YOUR OVERALL QUALITY OF LIFE?

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

Section 4.

This section asks how you *feel* about each of the following life areas.

Please circle the number that best describes how you **feel** about each area of your life.

---

1. I *feel* GUILTY

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

2. I *feel* FINANCIALLY DISADVANTAGED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

3. I *feel* ISOLATED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

4. I *feel* HOPE

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

5. I *feel* EXHAUSTED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

6. I *feel* SUPPORTED BY FAMILY AND FRIENDS

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

7. I *feel* SAD OR DEPRESSED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

8. I feel STRESSED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

9. I *feel* WORRIED THAT OUR FAMILY WILL NEVER BE FREE FROM HD BECAUSE OF ITS GENETIC NATURE.

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

10. I *feel* LIKE MY OWN NEEDS ARE NOT IMPORTANT TO OTHERS.

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

11. I *feel* COMFORTED BY THE BELIEF THAT ONE DAY THERE WILL BE A CURE FOR HD

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

12. In some ways I *feel* THAT HD HAS HAD A POSITIVE IMPACT ON MY LIFE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

13. I *feel* COMFORTED BY RELIGIOUS BELIEFS

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

14. I *feel* THAT I CAN COPE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

15. I *feel* THAT HD HAS MADE ME A STRONGER PERSON.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

16. I *feel* THAT I HAVE HAD A 'DUTY OF CARE' FORCED ON TO ME.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

17. I *feel* LIKE I DON'T KNOW WHO I AM ANYMORE.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

AND FINALLY, please tell us:

- **What you think would most improve your quality of life as a carer:**

- **Anything else related to your caring role that you feel hasn't been covered by this questionnaire.**

Thank you for your time.

Appendix VIII The Huntington's Disease Quality of Life Questionnaire for Carers - North American Version (HDQoL-C.US) - Pilot Version.

The Huntington's Disease Quality of Life Questionnaire for Carers (HDQoL-C) North American Pilot

Aimee Aubeeluck and Heather Buchanan

School of Education, Health and Sciences

UNIVERSITY of DERBY

Correspondence to:

Aimee Aubeeluck
Centre for Psychological Research in Health and Cognition
School of Education, Health Care and Sciences
University of Derby
Western Road
Mickleover DE3 9GX
Tel: 0870 040 7385
e-mail: a.aubeeluck@derby.ac.uk

Thank you for taking the time to fill in this questionnaire. The questionnaire has four sections. The first section will ask for some factual information. The next three will ask about different aspects of your role as a carer, how satisfied you are and how you feel about various aspects of your life.

We want to know how you feel about your quality of life, your health and other areas of your life. **Please answer all the questions.** If you are unsure about which response to give to a question, **please choose the ONE** that seems most appropriate (this is often your initial response).

Where there is an 11 point scale (0-10), please circle the number that you feel most accurately represents your situation. For example, a question might ask:

Q: Do you get the kind of support that you need from others?

Almost never								Almost always		
0	1	2	3	4	5	6	7	8	9	10

You should circle the number that best fits the kind of support you get. So if you very rarely get the kind of support from others that you need, you would circle the number 1.

Section 1.

This section asks for information about yourself. Please answer all the questions and do not spend too much time on any one item. The HD affected family member is sometimes referred to as a "person with HD" or simply pHD.

1a) What is your date of birth? ……..

b) What is your gender? (please circle) Male Female

c) How many years of education have you had? ……..

d) What is your marital status?

Single
Married
Living as married
Separated
Divorced

e) How long have you known about HD in your family? ……..

f) How long have you been caring for an HD affected family member? ……..

g) Are you the main carer for your HD affected family member? YES NO

h) What relation to you is the HD affected family member you are caring for? (e.g. spouse, sister, parent etc) ……..

i) Have you previously cared for any other HD affected family members? YES NO

- *if so*, what was their relationship to you? (e.g. spouse, sister, parent etc) ……..

j) Do you have any children? YES NO

- *if YES*, how many? ……..

How many are at risk?	How many have chosen to be tested for the HD gene?	How many have chosen NOT to be tested for the HD gene?

k) How many family members live in your household? ……..

2. **What is your personal or household (whichever is most relevant to you) gross annual income before tax?**

$_____

3. **Do you have any disabilities or medical conditions? (e.g. visual, hearing, physical, health, etc.).**

Yes ☐ No ☐

If yes please specify:

Name of disability or medical condition		*Extent of disability or medical condition*
e.g.	Visual	e.g. Require glasses for reading
	Diabetes mellitus	Require daily injections
	Epilepsy	Require daily medication
	Hypertension	Require daily medication
	High cholesterol	Require daily medication /change of diet
	Depression	Require daily medication
	Cardiac problems	Require daily medication
	Bone or muscle pain	Require daily medication
_____		_____
_____		_____
_____		_____
_____		_____

4. **How many hours do you spend on the following activities *each week*? (Average over past 3 months)**

Hours paid work outside the Home	0 ☐	1-10 ☐	11-20 ☐	21-30 ☐	31-40+ ☐
☐ urs unpaid child care	0	1-10	11-20 ☐	21-30 ☐	31-40+ ☐
☐ rs ur ☐ caring for pHD	0	1-10	11-20 ☐	21-30	31-40+ ☐
Hours paid to work caring for ☐ ☐	0	1-10	11-20	21-30	31-40+

Please specify any ***difficulties you experience*** caring for your pHD

e.g. Coping with behavior such as:	Irritability and temper outbursts lack of initiation Communication difficulties
Coping with physical problems such as:	Chorea Rigidity Swallowing difficulties
Coping with emotional problems such as:	Depression Mania Sexual disorders

Anything else?

_____

_____

_____

_____

_____

5. Is your home now suitable / suitably adapted for your family's needs?

Yes ☐ No ☐

If No please specify:

Area of concern	*Problem that it causes*
e.g. No stair lift No laundry room	e.g. Difficulty moving patient Difficulty getting the laundry done
_____	_____
_____	_____
_____	_____
_____	_____

6. **Below is a list of leisure activities. Indicate how often in an *average month* you attend, or take part, in each one of these leisure activities for your enjoyment (not employment).**

Activity		Number of times you do activity each month
(1)	Go to a club/group/society	_____
(2)	Go to a hotel/bar/pub	_____
(3)	Watch live sporting events (Not on TV)	_____
(4)	Go to a place of worship	_____
(5)	Chat with neighbours	_____
(6)	Eat out	_____
(7)	Go to the cinema	_____
(8)	Visit family or friend	_____
(9)	Play sport or go to a gym	_____
(10)	Other activities	_____

- Please tell us what these other activities are:

_____

_____

Section 2.

This section asks for information about different aspects of your role as a carer.
Please circle the number that you feel most accurately represents your situation. For example, a question might ask:

Q: Do you get the kind of support from others that you need?

Almost never — Almost always

0 1 2 3 4 5 6 7 8 9 10

You should **circle** the number that best fits the kind of support you get. So if you always get the kind of support from others that you need, you would circle the number 10.

---

1. How often are you restricted by the need to maintain a regimented daily routine?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

2. How often do you receive appropriate help from social services?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

3. How often do you have access to professionals who have specialised knowledge of HD and understand its implications?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

4. How much support are you given by health care professionals?

As much a I need — None Whatsoever

0 1 2 3 4 5 6 7 8 9 10

5. How often do the genetic implications of HD impact upon your caring role?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

6. How often do you have access to appropriate care facilities?

Whenever you need them — Almost never

0 1 2 3 4 5 6 7 8 9 10

7. How often do you receive any practical support that you need?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

8. How often do you experience a conflict of interest between what you want and what your HD affected relative wants?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

9. How often do you sleep well?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

Section 3.

This section asks how *satisfied* are you with each of the following life areas.

Please circle the number that best describes how **satisfied** you are with each area of your life.

1. **How *satisfied are you* with your HEALTH?**

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

2. **How *satisfied are you* with what you ACHIEVE IN LIFE?**

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

3. **How *satisfied are you with* your CLOSE RELATIONSHIPS WITH FAMILY OR FRIENDS?**

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

4. **How *satisfied are you* with HOW SAFE YOU FEEL?**

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

5. **How *satisfied are you* with FEELING A PART OF YOUR COMMUNITY?**

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

6. **How *satisfied are you* with YOUR OWN HAPPINESS?**

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

7. **How *satisfied are you* with THE TREATMENT THAT YOUR HD affected relative RECEIVES?**

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

8. **How *satisfied are you* with YOUR OVERALL QUALITY OF LIFE?**

Completely dissatisfied					Mixed					Completely satisfied
0	1	2	3	4	5	6	7	8	9	10

Section 4.

This section asks how you *feel* about each of the following life areas.

Please circle the number that best describes how you **feel** about each area of your life.

---

1. I *feel* GUILTY

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

2. I *feel* FINANCIALLY DISADVANTAGED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

3. I *feel* ISOLATED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

4. I *feel* HOPE

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

5. I *feel* EXHAUSTED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

6. I *feel* SUPPORTED BY FAMILY AND FRIENDS

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

7. I *feel* SAD OR DEPRESSED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

8. I feel STRESSED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

9. I *feel* WORRIED ABOUT THE GENETIC IMPLICATIONS OF HD

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

10. I *feel* LIKE MY OWN NEEDS ARE NOT IMPORTANT TO OTHERS.

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

11. I *feel* COMFORTED BY THE BELIEF THAT ONE DAY THERE WILL BE A CURE FOR HD

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

12. In some ways I *feel* THAT HD HAS HAD A POSITIVE IMPACT ON MY LIFE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

13. I *feel* COMFORTED BY RELIGIOUS BELIEFS

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

14. I *feel* THAT I CAN COPE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

15. I *feel* THAT HD HAS MADE ME A STRONGER PERSON.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

16. I *feel* THAT I HAVE HAD A 'DUTY OF CARE' FORCED ON TO ME.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

17. I *feel* LIKE I DON'T KNOW WHO I AM ANYMORE.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

AND FINALLY, please tell us:

- What you think would most improve your quality of life as a carer:

- Anything else related to your caring role that you feel hasn't been covered by this questionnaire.

Thank you for your time.

Appendix IX The Huntington's Disease Quality of Life Questionnaire for Carers (HDQoL-C) - User Manual (© Aubeeluck & Buchanan, 2004).

Huntington's Disease Quality of Life Battery for Carers.

(HDQoL-C)

Validated for use with Spousal Carers of Persons with Huntington's Disease.

Aimee Aubeeluck and Heather Buchanan

School of Education, Health and Sciences
University of Derby

UNIVERSITY of DERBY

Correspondence to:

Aimee Aubeeluck
Centre for Psychological Research in Health and Cognition
School of Education, Health Care and Sciences
University of Derby
Western Road
Mickleover DE3 9GX
Tel: 0870 040 7385
e-mail: a.aubeeluck@derby.ac.uk

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1 Introduction

1.1 Defining Quality of life

The World Health Organisation (WHO) define Quality of Life as,

"... .an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns". (WHO Group, 1995, pg 3).

This is a broad ranging concept which incorporates the individual's physical health, psychological state, level of independence, social relationships, personal beliefs and relationship to salient features of the environment. Furthermore, this definition highlights the view that quality of life refers to a subjective evaluation, which includes both positive and negative dimensions, and which is embedded in a cultural, social and environmental context. The scale that follows is an operationalization of the WHO's definition of quality of life.

1.2 Quality of life as an outcome measure.

Since the 1960's QoL has been emerging as a useful outcome measure by which to judge the efficacy of psychological interventions (e.g. Cummins, 1997; Rapley, 2003). Using QoL as a measure of outcome focuses on the impact of a condition or situation on the *individual's* emotional and physical functioning and lifestyle. As such, QoL indicators can assess the subjective benefit of interventions and help to answer the question of whether an intervention leads to an increase in wellbeing by providing a client-led baseline against which its effects can be evaluated (Bowling, 2001).

1.3 Objective of the HDQoL-C.

The objective of the HDQoL-C is to quantify the caregiving experience in Huntington's Disease (HD). in order to implement and assess interventions.

The HDQoL-C:

(a) **Is Multidimensional.** The HDQoL-C incorporates the individual's physical health, psychological state, level of independence, social relationships, personal beliefs and relationship to salient features of the environment.

(b) **Measures QoL from a subjective perspective.** The HDQoL-C has been developed primarily as an outcome measure i.e. to assess the efficacy of interventions by providing a baseline from which the impact of an intervention can be measured. As such, it does not use the scores obtained as a specific indicator of the QoL that each client is experiencing, but merely as a measure by which to assess whether the client perceives their QoL to be increasing or decreasing when reassessed at a later stage.

(c) **Has been validated for use with SPOUSAL carers of HD patients.** The symptoms and genetic nature of HD makes the spousal carer role distinct from other HD family carer roles (e.g Kessler, 1993). At this present time, the HDQoL-C has only been validated for use with HD spousal carers. However, it is thought that the HDQoL-C may also prove useful in assessing the QoL of other HD family carers, (e.g. child carers, carers who are at risk or carers who are HD positive) and research is ongoing with regards to these specific populations. Therefore, any client assessments / research findings that are obtained using the HDQoL-C with such populations should be interpreted with caution.

(d) **Is psychometrically sound.** The HDQoL-C is adapted from the Comprehensive Quality of life Scale for Adults (COMQoL-A5, Cummins, 1997), a well validated and documented QoL tool designed for use with the general adult population. The HDQoL-C demonstrates good internal consistency, test re-test reliability and congruent validity (see section 5). However, it should be noted that the HDQoL-C has been validated using a UK population of spousal carers. It is therefore advisable for the researcher to check the psychometric properties e.g., Cronbach's alpha coefficient if the scale is used outside the UK and / or with a different carer population.

2. Administration.

2.1 General information

The scale is intended to be self-administered.

It should be noted that the instrument exists in four parts:
1) Demographic / objective Information
2) Aspects of caring
3) Satisfaction with life
4) Feelings about life

Component 1 requests demographic and objective information from the client and each question is treated independently. This information can be used in research to investigate the factors that may predict QoL in caregiving. Alternatively, it may be used by the practitioner to build up an overall picture of a client. As this component does not in itself constitute a scale, the researcher / practitioner is able to omit questions or include additional questions that may be of interest in this section.

Components 2, 3 and 4 all comprise differing aspects of disease-specific and subjective QoL. Each component shows good internal consistency, test-retest reliability and congruent validity (see section 6). Moreover, each component demonstrates a moderate to strong correlation with the others allowing researchers / practitioners to use any combination or all of the components to investigative either specific issues surrounding the QoL of the HD-spousal carer or overall QoL scores.

The HDQoL-C takes about 10-20 minutes to complete.

The Huntington's Disease Quality of Life Battery for Carers (HDQoL-C)

Aimee Aubeeluck and Heather Buchanan

School of Education, Health and Sciences

UNIVERSITY of DERBY

Correspondence to:

Aimee Aubeeluck
Centre for Psychological Research in Health and Cognition
School of Education, Health Care and Sciences
University of Derby
Western Road
Mickleover DE3 9GX
Tel: 0870 040 7385
e-mail: a.aubeeluck@derby.ac.uk

Thank you for taking the time to fill in this questionnaire. The questionnaire has four sections. The first section will ask for some factual information. The next three will ask about different aspects of your role as a carer, how satisfied you are and how you feel about various aspects of your life.

We want to know how you feel about your quality of life, your health and other areas of your life. **Please answer all the questions.** If you are unsure about which response to give to a question, **please choose the ONE** that seems most appropriate (this is often your initial response).

Where there is an 11 point scale (0-10), please circle the number that you feel most accurately represents your situation. For example, a question might ask:

Q: Do you get the kind of support from others that you need?

Almost never								Almost always		
0	1	2	3	4	5	6	7	8	9	10

You should **circle** the number that best fits the kind of support you get. So if you very rarely get the kind of support from others that you need, you would circle the number 1.

Section 1.

This section asks for information about yourself. Please answer all the questions and do not spend too much time on any one item.

1a) What is your date of birth? ... ...

b) What is your sex ? (please circle) Male Female

c) What is the highest education you received?
Primary school
Secondary school
University
Post-graduate

d) What is your marital status?
Single
Married
Living as married
Separated
Divorced

e) How long have you known of the presence of HD in your family? ... ...

f) How long have you been caring for an HD affected family member? ... ...

g) Are you the main carer for your HD affected family member? YES NO

h) What relation to you is the HD affected family member you are caring for? (e.g. spouse, sister, parent etc) ... ...

i) Have you previously cared for any other HD affected family members? YES NO

- *if so*, what was their relationship to you? (e.g. spouse, sister, parent etc) ... ...

j) Do you have any children? YES NO
- *if so*, what was is their genetic status? (e.g. HD positive/ negative, 'at-risk' but not tested) ... ...

2. **What is your personal or household (whichever is most relevant to you) gross annual income before tax?**

£_____

3. **Do you have any disabilities or medical conditions? (e.g. visual, hearing, physical, health, etc.).**

Yes ☐ No ☐

If yes please specify:

Name of disability or medical condition	*Extent of disability or medical condition*
e.g. Visual	e.g. Require glasses for reading
Diabetes	Require daily injections
Epilepsy	Requires daily medication
_____	_____
_____	_____

4. **How many hours do you spend on the following *each week*? (Average over past 3 months)**

Hours paid work	0	☐	1-10	☐	11-20	☐	21-30	☐	31-40+ ☐
☐ urs u☐ child care	0		1-10		11-20	☐	21-30	☐	31-40+ ☐
☐ urs u☐ caring for pHD	0		1-10		11-20	☐	21-30		31-40+ ☐

Please specify any ***difficulties you experience*** caring for your person with Huntington's Disease (pHD) (e.g. dealing with behaviour, physical problems, emotional problems etc)

_____

_____

_____

5. **Is your home suitable / suitably adapted for your family's needs?**

Yes ☐ No ☐

If No please specify:

Area of concern	*Problem that it causes*
e.g. No stair lift	e.g. Difficulty moving patient
No utility room	Difficulty getting the laundry done
_____	_____
_____	_____

6. **Below is a list of leisure activities. Indicate how often in an *average month* you attend, or take part in, each one for your enjoyment (not employment).**

Activity		Number of times you do activity each month
(1)	Go to a club/group/society	_____
(2)	Go to a hotel/bar/pub	_____
(3)	Watch live sporting events (Not on TV)	_____
(4)	Go to a place of worship	_____
(5)	Chat with neighbours	_____
(6)	Eat out	_____
(7)	Go to the cinema	_____
(8)	Visit family or friend	_____
(9)	Play sport or go to a gym	_____
(10)	Other activities	_____

- Please tell us what these other activities are:

_____

_____

Section 2.

This section asks for information about different aspects of your role as a carer.
Please circle the number that you feel most accurately represents your situation. For example, a question might ask:

Q: Do you get the kind of support from others that you need?

Almost never — Almost always

0 1 2 3 4 5 6 7 8 9 10

You should circle the number that best fits the kind of support you get. So if you always get the kind of support from others that you need, you would circle the number 10.

---

1. How often are you restricted by the need to maintain a regimented daily routine?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

2. How often do you receive appropriate help from social services?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

3. How often do you have access to professionals who have specialised knowledge of HD and understand its implications?

Whenever I need it — Almost never

0 1 2 3 4 5 6 7 8 9 10

4. How much support are you given by health care professionals?

As much a I need — None Whatsoever

0 1 2 3 4 5 6 7 8 9 10

5. How often do the genetic implications of HD impact upon your caring role?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

6. How often do you have access to appropriate care facilities?

Whenever you need them — Almost never

0 1 2 3 4 5 6 7 8 9 10

7. How often do you receive any practical support that you need?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

8. How often do you experience a conflict of interest between what you want and what your HD affected relative wants?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

9. How often do you sleep well?

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

Section 3.

This section asks how *satisfied* are you with each of the following life areas.

Please circle the number that best describes how **satisfied** you are with each area of your life.

---

1. How *satisfied are you* with your HEALTH?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

2. How *satisfied are you* with what you ACHIEVE IN LIFE?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

3. How *satisfied are you with* your CLOSE RELATIONSHIPS WITH FAMILY OR FRIENDS?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

4. How *satisfied are you* with HOW SAFE YOU FEEL?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

5. How *satisfied are you* with FEELING A PART OF YOUR COMMUNITY?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

6. How *satisfied are you* with YOUR OWN HAPPINESS?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

7. How *satisfied are you* with THE TREATMENT THAT YOUR HD affected relative RECEIVES?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

8. How *satisfied are you* with YOUR OVERALL QUALITY OF LIFE?

Completely dissatisfied — Mixed — Completely satisfied

0 1 2 3 4 5 6 7 8 9 10

Section 4.

This section asks how you *feel* about each of the following life areas.

Please circle the number that best describes how you **feel** about each area of your life.

---

1. I *feel* GUILTY

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

2. I *feel* FINANCIALLY DISADVANTAGED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

3. I *feel* ISOLATED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

4. I *feel* HOPE

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

5. I *feel* EXHAUSTED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

6. I *feel* SUPPORTED BY FAMILY AND FRIENDS

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

7. I *feel* SAD OR DEPRESSED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

8. I feel STRESSED

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

9. I *feel* WORRIED ABOUT THE GENETIC IMPLICATIONS OF HD

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

10. I *feel* LIKE MY OWN NEEDS ARE NOT IMPORTANT TO OTHERS.

Almost always 0 1 2 3 4 5 6 7 8 9 10 Almost never

11. I *feel* COMFORTED BY THE BELIEF THAT ONE DAY THERE WILL BE A CURE FOR HD

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

12. In some ways I *feel* THAT HD HAS HAD A POSITIVE IMPACT ON MY LIFE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

13. I *feel* COMFORTED BY RELIGIOUS BELIEFS

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

14. I *feel* THAT I CAN COPE

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

15. I *feel* THAT HD HAS MADE ME A STRONGER PERSON.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

16. I *feel* THAT I HAVE HAD A 'DUTY OF CARE' FORCED ON TO ME.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

17. I *feel* LIKE I DON'T KNOW WHO I AM ANYMORE.

Almost always — Almost never

0 1 2 3 4 5 6 7 8 9 10

AND FINALLY, please tell us:

- **What you think would most improve your quality of life as a carer:**

- **Anything else related to your caring role that you feel hasn't been covered by this questionnaire.**

Thank you for your time.

4.1 Forms of data analysis

4.1.1 For the practitioner or service provider

The most useful level of analysis may be in terms of component scores rather than an overall QoL score. As such, each area where life quality may be suffering can be monitored individually in line with interventions.

For component 1: These are demographic and objective variables. They do not need to be totaled in any way but can be used to build up an overall picture of the client.

For component 2: Each of the 9 scores can simply be added up to give a Total score

Note:
Items 2, 3, 4, 6, 7,and 9 are all positively worded and therefore need reversing before scores are totaled (see section 4.2.2 for further information).

Alternatively if you prefer to work in percentages, use the formula:
[(N/90)*100 = % Aspects of caring score].

A 100% score would reflect an optimum integration with the caring role with very little impact of carer role on the QoL of the client.

For component 3:

Each of the 8 scores can simply be added up to give a Total score.

Alternatively if you prefer to work in percentages, use the formula:
[(N/80)*100 = % Satisfaction with life].

A 100% score would reflect optimum satisfaction with life and represent an optimum QoL.

For component 4:

Each of the 17 scores can simply be added up to give a Total score.

Note:
Items 4, 6, 11, 12, 13, 14 and 15 are all positively worded and therefore need reversing before scores are totaled (see section 4.2.4 for further information).

Alternatively if you prefer to work in percentages, use the formula:
[(N/170)*100 = % Satisfaction with life].

A 100% score would reflect optimum feelings about life and is strongly correlated with optimum QoL.

Note
Make sure all scores that need to be reversed are altered before the scores are totaled.

4.1.2 For the researcher

The most useful level of analysis may be the overall QoL scores obtained.

To obtain the overall QoL score: follow section 4.5.1. guidelines for practitioners or service providers, then, to calculate the overall QoL score, use the formula:

[(% component 2 + % component 3 + % component 4)/ 3 = % overall QoL score]

4.1.3 Why calculate the percentage scores?

The HDQoL-C has been developed primarily as an outcome measure i.e. to assess the efficacy of interventions by providing a baseline from which the impact of an intervention can be measured. As such, it does not use the scores obtained as a specific indicator of the QoL that each client is experiencing, but merely as a measure by which to assess whether the client perceives their QoL to be increasing or decreasing when reassessed at a later stage. Therefore, using a lengthy formula is unnecessary and totalling the scores (although this is perfectly acceptable) will only provide abstract numbers for the practitioner or researcher to work with. By calculating percentage scores, the QoL battery becomes an understandable gradient of percentage scores for practitioners and researchers alike to use.

4.2 Entering Data into a statistical package.

If you are using a statistical package such as SPSS, enter the items using the following procedure:

4.2.1 Demographic and Objective Information

This information can be used in research to investigate the factors that may predict QoL in spousal caring. Alternatively, it can be used by practitioners to gain an overall picture of a client.

Note:
Each item in this component should be treated independently i.e. the questions do not constitute a scale.

1a) What is your date of birth?
Enter date of birth in months or years

b) What is your sex ? (please circle)
Enter 1 for Male and 2 for Female

c) What is the highest education you received?
Enter 1 for Primary school
2 for Secondary school
3 for University
4 for Post-graduate

d) What is your marital status?
Enter 1 for Single
2 for Married
3 for Living as married
4 for Separated
5 for Divorced

e) How long have you known of the presence of HD in your family?
Enter time in months or years

f) How long have you been caring for an HD affected family member?
Enter time in months or years (be consistent with question 1e)

g) Are you the main carer for your HD affected family member?
Enter 1 for yes and 2 for no

h) What relation to you is the HD affected family member you are caring for? (e.g. spouse, sister, parent etc)
Enter 1 for spouse. If you are using the questionnaire with a different population, please code as necessary.

i) Have you previously cared for any other HD affected family members?
Enter 1 for yes and 2 for no
- *if so*, what was their relationship to you?

Code this as necessary (e.g. 1 for sibling, 2 for parent, 3 for friend etc)

j) Do you have any children?
Enter 1 for yes and 2 for no
- *if so*, what was is their genetic status?

Code this as necessary (e.g. 1 for HD positive, 2 for at risk, 3 for negative etc)

2) What is your personal or household (whichever is most relevant to you) gross annual income before tax?
Enter income.

3) Do you have any disabilities or medical conditions? (e.g. visual, hearing, physical, health, etc.).
Enter 1 for yes and 2 for no
***For research purposes, you may want to code disabilities / medical conditions in terms of severity e.g.**

5 = No disability
4 = Minor disability (e.g. eyeglasses) not likely to interfere with normal life activities or routines
3 = Constant, chronic condition that interferes to some extent with daily life (eg. diabetes, heart condition, migraines, infertility, asthma.
2 = Disability likely to restrict social activities (e.g. profound deafness, blindness, significant physical disability, depression, arthritis, asthma needing regular medication)
1 = Major disability likely to require daily assistance with personal care (e.g. severe psychiatric condition, advanced multiple sclerosis, severe cognitive or physical impairment, quadriplegia) (coding taken from Cummins 1997)

Note
It is sometimes difficult to choose between categories, but as long as you are consistent and make note of your classifications, this is fine.

4) How many hours do you spend on the following *each week*? (Average over past 3 months)
For each type of work, enter:
1 for 31-40+ hours
2 for 21-30 hours
3 for 11 - 20 hours
4 for 1 - 10 hours
5 for none

- Please specify any **difficulties you experience** caring for your pHD: (e.g. dealing with behaviour, physical problems, emotional problems etc).
  Researchers may want to code these difficulties.
  It may be more useful for practitioners just to make note of them.

5) Is your home suitable / suitably adapted for your family's needs?
Enter 1 for yes and 2 for no

- If No please specify:
  Researchers may want to code these difficulties.
  It may be more useful for practitioners just to make note of them.

6) Below is a list of leisure activities. Indicate how often in an *average month* you attend or do each one for your enjoyment (not employment).
Total and Enter the number of times client has carried out an activity.
Note: the amount of times a client chats with neighbours may inflate the score considerably giving an unrealistic picture of social activity.

4.2.2 Aspects of caring

Some of the questions in this section will need to reversed before analysis. However, it is suggested that the data are entered into the computer as circled, and that subsequent re-coding takes place within the computer. Consequently, data should be entered as follows:

For each of the 9 questions, simply enter the number that the client has entered.

The following questions are worded positively and as such need reversing in order that a negative response does not give a positive score (it is advisable to use your statistical package to do this although it can be done by hand).

2. How often do you receive appropriate help from social services?
3. How often do you have access to professionals who have specialised knowledge of HD and understand its implications?
4. How much support are you given by health care professionals?
6. How often do you have access to appropriate care facilities?
7. How often do you receive any practical support that you need?
9. How often do you sleep well?

Therefore, for each of these questions an answer of:
0 is reversed to 10
1 is reversed to 9
2 is reversed to 8
3 is reversed to 7
4 is reversed to 6
5 is reversed to 5
6 is reversed to 4
7 is reversed to 3
8 is reversed to 2
9 is reversed to 1
10 is reversed to 0

4.2.3 Satisfaction with life

For each of the 8 questions, simply enter the number that the client has entered.

4.2.3 Feelings about life.

For each of the 17 questions, enter the number that the client has entered.

The following questions are worded positively and as such need reversing.

4. I *feel* HOPE
6. I *feel* SUPPORTED BY FAMILY AND FRIENDS
11. I *feel* COMFORTED BY THE BELIEF THAT ONE DAY THERE WILL BE A CURE FOR HD
12. In some ways I *feel* THAT HD HAS HAD A POSITIVE IMPACT ON MY LIFE
13. I *feel* COMFORTED BY RELIGIOUS BELIEFS
14. I *feel* THAT I CAN COPE
15. I *feel* THAT HD HAS MADE ME A STRONGER PERSON.

Reverse as in section 4.2.2

Note
Use the score of 99 to allow computer identification of missing values. If this scheme is used, care needs to be taken that these '99' values are recognized as excluded values, and not included as data.

4.3 Data cleaning

It is recommended that the raw data files be carefully examined prior to the implementation of analytic procedures. In particular;

(a) Analysis of response frequency data for each variable will allow the researcher to establish that the computer is recognising '99' as a missing variable.
(b) The raw data for each client can also be visually scanned to detect patterns of response that are consistently at the top or bottom of the likert scales. Such data should be excluded prior to analysis since they provide no variance and are likely to reflect floor and ceiling effects.

4.4 Dealing with a data skew

You may have data that is moderately negatively skewed. To restore normality, a square root transformation can be used. However, opinion is divided among statisticians as to whether this procedure is appropriate. In line with Tabachnick and Fidell (1996), it is **not** recommended that the data be transformed for the HDQoL-C.

4.5 Why use an 11 point likert scale?

A major problem with QOL data is their tendency to cluster at the favorable end of any scale (Cummins, 1997). Furthermore, likert scales are often criticized for being unnecessarily restrictive (Fayers & Machin, 2001). Therefore, utilizing an 11 point response should make the Likert methodology more successful as it allows for a wide range of scale levels.

5.1 Construction of the HDQoL-C

The HDQoL-C is based upon the domains and facets of the Comprehensive Quality of Life scale for adults (ComQol-A5, Cummins 1997). A well validated and documented QoL tool designed for use with the general adult population.

5.2 Validity:

5.2.1. *Congruent Validity:*

Congruent validity establishes the validity of a new test by correlating scores from it with scores from another test with established validity. The HDQoL-C was correlated with the WHOQOL-BREF (WHO, 1996), a well documented and validated QoL measurement for use with the general adult population.. The relationship between the scores on the HDQoL-C and the WHOQOL-BREF revealed a Pearson's correlation coefficient of r=0.58 for component 2 (Aspects of caring), r= 0.64 for component 3 (satisfaction with life and r=0.76 for component 3 (Feelings about life). This moderate to strong association with WHOQOL-BREF confirms that the HDQoL-C is measuring QoL.

Each component further correlates with the other two components of the HDQoL-C as follows:

	Component 1	Component 2	Component 3
Component 2	1.00	0.56	0.51
Component 3	0.56	1.00	0.83
Component 4	0.51	0.83	1.00

Note:

Component 1 is demographical / objective data and each question is treated independently. Therefore, this component cannot be correlated with either the WHO-BREF or components 2, 3, and 4 of the HDQoL -C.

5.3 Reliability:

5.31. Internal consistency:

Internal consistency (components 2. 3 and 4) was assessed using Cronbach's Alpha coefficient, which measures the overall correlation between items as well as the level of correlation between items within a scale. Reliability estimates of 0.7 and 0.9 are recommended for instruments that are used in groups and individuals respectively.

	Cronbach's alpha
Component 2	0.801
Component 3	0.844
Component 4	0.885

5.3.2 Test-retest reliability:

Reproducibility assesses whether an instrument produces the same results on repeated administrations when respondents have not changed. This is assessed by test-retest reliability. The reliability coefficient was calculated by correlating instrument scores for the two administrations. Thus, 10 carers completed the HDQoL-C two weeks after first administration. Both questionnaire scores were subsequently correlated using Pearsons correlation coefficient.

	Pearsons correlation (r=)
Component 2	0.86
Component 3	0.90
Component 4	0.92

Note:

Please contact corresponding author for any further details of item analysis if required. We are continuing to develop and standardise the HDQoL-C for use in Britain and globally. It would be most helpful if researchers could send copies of any raw data gathered using the HDQoL-C to the corresponding author on diskette or electronically. This will enable us to continue to develop the instrument at national and international levels. Data contributions to any publications will be acknowledged in the usual way.

6 References

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