

HUNTINGTON'S DISEASE: TIME TO CARE

HUNTINGTON'S DISEASE: TIME TO CARE

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ABSTRACT

Huntington's disease (HD) is a devastating, degenerative brain disorder for which there is, at present, no effective treatment or cure. HD slowly diminishes the affected individual's ability to walk, think, talk and reason (Huntington's Disease Society of America, 2007).

Huntington's disease profoundly affects the lives of entire families – emotionally, socially and economically. Despite this impact, the predominant research focus is on finding a cure and the medical treatment of the disease. There has been very little research on the meaning of the illness experience for individuals with HD and families.

This study explored six people's experiences, two people with HD, and four family-care providers, with the intent of representing their experience as intimately as possible. The following themes emerged from the analysis of the participants' narratives: the challenge to social identity; efforts to sustain social identity; and the mismatch between the needs of people with HD and the available formal care.

Family members emphasized certain characteristics of good care: empathic, flexible, accessible and responsive to individual and family needs. Families in this study identified the moral dilemma they were faced with knowing that the care their relative with HD received was inadequate, yet there was limited community support such as home care to augment the significant emotional and physical demands associated with caring for the person with HD at home.

Family members valued empathic care providers who recognized the person with HD was not culpable for his or her behaviour. Thus the provision of care requires better training, and institutional arrangements that allow for the time and flexibility that the person with HD requires.

It is also apparent from this study that family members and individuals with HD can make a significant contribution to understanding the social and emotional implications of the illness.

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INTRODUCTION

The Huntington's Disease Society of America (HDSA) describes Huntington's disease (HD) as a devastating, degenerative brain disorder for which there is, at present, no effective treatment or cure. HD slowly diminishes the affected individual's ability to walk, think, talk and reason (HDSA, 2007). Symptoms progressively worsen over a 10 to 20 year period until the person with HD becomes totally dependent on others for his or her care. Some symptoms such as choreic movements and mood swings differ between individuals even within the same family, thus care must be specialized to meet individual needs.

It is estimated that one in every 10,000 Canadians has Huntington's disease and five in every 10,000 is at risk of developing the disease (Huntington Society of Canada [HSC], 2007). Males and females of all ethnic origins are affected equally, and each child of a person with HD has a 50 percent chance of inheriting the gene.

Huntington's disease profoundly affects the lives of entire families – emotionally, socially and economically. Completed suicide is as high as 13% in HD, with suicidal ideation occurring in up to 50% in those with the disease (Paulsen, Hoth, Nehl, Stierman, 2005; Wahlin, 2000). Despite the social impact of the illness on individuals and families the predominant research focus is on finding a cure and the medical treatment of the disease.

This research focuses on the lived experience of Huntington's disease for individuals with HD and families. My purpose is to represent their experiences as closely

as possible in order to highlight the tremendous social impact of the disease on peoples' lives, and the inadequate care and supports available to buffer the ravages of the illness. Despite the abiding strengths and support of family in sustaining their family member with HD, the illness context is one of isolation, and often despair. It is apparent from this study that informed, responsive, compassionate care could help to sustain the individual with HD and family through the debilitating course of the illness.

To provide context for the research, I offer first an overview of the illness and then a discussion of the historical treatment of people with Huntington's. This is followed by a discussion of the health system context. I then review the literature on the social and lived aspects of Huntington's disease.

Context for the Research

The Illness

Multiple physical, cognitive and emotional changes are part of Huntington's disease. The chorea associated with HD results in individuals walking with an unusual gait, usually with writhing dance like movements. The chorea also causes problems with feeding oneself along with the potential risk of choking and falls as the illness progresses.

People with Huntington's develop communication difficulties such as delayed response and reduced speech and phrases. In the mid-stages of HD, people lose precision in making sounds, control of the volume of sounds they make, and coordination of speech and breathing mechanisms (Sullivan, Bird, Alpay, Cha, 2001). Speech at this stage may vary in volume, be interrupted by grunting or heavy breathing and hard to understand.

The ability to communicate worsens as the disease progresses. In the advanced stage of the illness, people have only a few intelligible words or sounds to express their range of needs and emotions (Pollard, Best, Imbriglio, Klasner, Rubin, Sanders, Simpson, 1999). These changes have been attributed to damage to specific parts of the brain. This inability to express one's thoughts and feelings can contribute to the individual's frustration and the triggering of emotional outbursts (Sullivan, Bird, Alpay, Cha, 2001). It also contributes to misconceptions about the individual's competency and misunderstandings on the part of care-providers (Sullivan et al, 2001).

Some health professionals view people with HD as impaired in all functions. Snowden (2001) proposes that the belief that the person with HD is globally impaired is misleading and inaccurate. Studies have shown that HD damages selective parts of the brain, yet other parts continue to function well (Snowden, 2001). A four-year study of cognitive functioning in Huntington's disease found only modest decline on most neuropsychological variables (Ward, Sheppard, Shpritz, Margolis, Rosenblatt & Brandt, 2006). Some aspects of the individual's intellectual functioning remain well-preserved, such as the ability to see and hear and understand the meaning of what they see and hear, even when the illness is advanced (Snowden, 2001; Sullivan et al 2001; Goolkasian; 2001; Shannon, 2006; Ball, 1982).

The assumption that the person with HD experiences global impairment similar to Alzheimer's disease has a dramatic impact on the care that the individual receives by limiting further exploration and understanding of other features of the illness. This assumption has been challenged in the past (Chiu, 1989a; Ball, 1982) and continues to be

at present (Snowden, 2001; Moskowitz, Marder, 2001; National Institute of Neurological Disorders and Stroke, 2007, Shannon, 2006) yet also continues to be represented in current academic literature (Blass, Steinberg, Leroi &Lyketsos, 2001).

The specific difficulties in thinking associated with HD give rise to specific and predictable changes in behaviour. Changes in behaviour are a central feature of Huntington's disease, and are often identified as the most distressing aspects of the condition that create the greatest challenge for care-providers (Snowden, 2001). Individuals with HD often have attention overload in attempting to carry out activities that are generally taken for granted by most able-bodied people such as walking and talking. The conscious effort required to perform any activity including switching attention from one task to another, such as responding to a question while watching television, requires extreme effort (Pollard, Best, Imbriglio, Klasner, Rubin, Sanders, Simpson, 2003).

The History of Huntington's disease

Reflecting on the history of Huntington's disease provides context to the social impact of the illness, and is in keeping with this study's methodology of interpretive phenomenology, which in part draws its understanding of meanings through historical changes (Benner, 1994). The history of HD is a history of emerging medical knowledge and social advocacy, and also of marginalization and violence.

It is thought that HD has been in existence since the 1600's and that many of the witches burned at the stake in Salem Massachusetts were actually people with Huntington's disease (Hayden, 1983, as cited in Cox, 1999) The symptoms of the illness,

that is the dance-like uncoordinated movements (chorea) and odd behaviour were perceived as possession by the devil (Chen, 2002, Schiel, 2004). Physicians in the United States England and Norway in the 1840's documented accounts of people with involuntary movements and "mental disturbances" that were inherited from a similarly-affected parent, and HD was described in the medical literature as "chronic hereditary chorea"(Chen,2002, Schiel,2004). In 1872 George Huntington wrote an influential paper entitled "On Chorea," using personal accounts of his father's patients and in response the disease was named after him (Cox,1999).

In the 1900's Charles Davenport, a eugenicist, argued for compulsory sterilization and immigration restriction for those afflicted with HD (Chen,2002, Schiel, 2004). Along with other social groups seen as defective, individuals with HD were targeted and put to death by the Nazis during the Second World War.

With the 1953 discovery of DNA's structure by Watson and Crick there was an increase in publications on HD research (Chen, 2002, Schiel, 2004). In 1981 Nancy Wexler, an American psychologist, whose mother had died of HD, began fieldwork in Lake Maracaibo, Venezuela, an area with unusually high numbers of individuals affected by HD. This research led to the discovery of the Huntington's gene in 1993. Genetic testing confirms the Huntington's gene but not at what point the disease will occur. Since at present there is no effective treatment or cure for HD, genetic testing may add to the emotional burden experienced by individuals and families (HDSA, 2007)

In 1966 Myrianthopoulous spoke out about the lack of knowledge of HD. This lack of knowledge was highlighted with the death in 1967 of Woody Guthrie the famous

folk singer who wrote “This Land Is Your Land”. He died of HD in the back ward of a psychiatric institution in New York City having been diagnosed initially with schizophrenia and alcoholism (Chiu, 1989b). His wife, Marjorie, driven by the lack of research interest in Huntington’s disease and support for families, started what is now called the Huntington’s Disease Society of America to provide public health research on HD. There are now chapters across Canada and around the world.

In 1972, an International Centennial Symposium on Huntington’s was held which gathered all HD researchers in an attempt to assess what was known about HD, and spur new interest in research. Milton Wexler, a prominent psychoanalyst whose wife had died of HD in 1978, established the Hereditary Disease Foundation, a think tank to encourage open dialogue among researchers in the pursuit of a cure (Hereditary Disease Foundation, 2007). In the late 70’s researchers found evidence of HD affecting cells all over the body, not just the brain.

The bibliography on Huntington’s disease is vast and has expanded in the past decade (Hereditary Disease Foundation, 2007). PubMed, a service of the National Library of Medicine, provides access to over 12 million Medline citations back to the mid-1960’s and additional life science journals (Hereditary Disease Foundation, 2007).

In 2006, Dr. Michael Hayden a Canadian researcher and his team, provided evidence for a cure for Huntington’s disease in a mouse, offering hope that the disease can be relieved in humans (Hereditary Disease Foundation, Research Updates, 2006).

Dr. Edmond Chiu, Honorary Professorial Fellow in Psychiatry at the University of Melbourne was a guiding force in the opening of the Arthur Preston Centre, a

specialized care centre for people with HD in Balwyn, Australia in 1981 (Ball, 1982).

Despite this example of a care facility focused on the quality of life for those with HD, there are few care facilities that specifically address the unique needs of people with HD in North America (Smith, 2002).

The Health System Context

The social construction of Canadian health care reflects the central positioning of the biomedical model and the resulting medicalization of health and issues related to health care (Penning, 2002). In this context, health tends to be defined using biomedical criteria and is conceptualized as the absence of disease or pathology (Penning, 2002). These perspectives influence what is seen as the primary focus of the health care system, the treatment and cure of acute conditions by physicians, largely within acute-care hospital settings (Penning, 2002). It follows that individuals with chronic disease, including of course Huntington's disease, remain marginal to this system.

Strategies to reform the health care system are focused mainly on cutting costs and community-based care services such as home care are promoted as a less expensive alternative to acute and long-term hospitals and institutional care (Taylor, 1990; Williams 1996 as cited in Penning, 2002). Reduced health care funding has resulted in hospital and hospital bed closures, declines in admission rates, and shortened hospital stays. However, these changes have not been paralleled with additional resources to the community care sector that would shore up as well as develop new supportive services (Chappell, 1995 as cited in Penning, 2002; Skirton and Glendinning, 1997; Aronson, 2002).

Research evidence further suggests that institutional and community settings provide only a minimal amount of the care that people receive and the majority of chronic and long-term care continues to be self-administered, or provided by family, friends or others in the community (Levin & Idler, 1981 as cited in Penning, 2002). Family members are the major source of informal care and provide an estimated 75 – 80% of the long-term care received (Chappell, 1994; Marshall, 1994, as cited in Penning, 2002). Long-term and chronic care is thus structured as a private responsibility to be assumed by individuals and families (Penning, 2002). People with Huntington's disease, and their families, like so many others facing long-term and chronic illnesses, often bear the full weight of care (Snowden, 2001; Aronson, 2002; Aubeeluck & Buchanan, 2006; Yale & Martindale, 1984; Dawson, Kristjanson, Toye & Flett, 2004). A study undertaken by Ohman and Soderberg (2004) probed the meaning of close relatives' experiences of living with a person with chronic illness. Their findings suggest that chronic illness has a direct impact on the life course of healthy relatives, inducing feelings of powerlessness, distress, restriction and alienation, and a lack of sufficient support from formal health care providers to relatives despite this knowledge.

LITERATURE REVIEW

There is very little written from a social science perspective on Huntington's disease. A search of sociological abstracts and social service abstracts for Huntington's disease as a keyword, turned up 22 peer-reviewed articles. Of these, the vast majority tended to focus on the ethical and moral implications of genetic testing. Only three articles explored issues relevant to this thesis. Miller (1976) Sands (1984) Yale & Martindale (1984) explore the impact of family involvement and support for the individuals with HD, as well as the role social work can play in enhancing the strength of families.

Parker (1994) compiled a sociological personal account of the negligent, often abusive care his wife who had Huntington's disease received in a series of nursing homes. He then contrasted this experience to the specialized care she received at a 'model' residential care facility devoted specifically to Huntington's disease patients. Miller (1976) highlighted the work of the Delaware Valley chapter of the Committee to Combat Huntington's Disease [CCHD]. Their underlying goal was to bring strength to families under stress due to Huntington's disease. They developed insights and perspectives on the disease by listening to families articulate their needs. They then developed specific services in conjunction with families that were flexible, and respected individual family needs and strengths. Sands (1984) outlined the specialized role for social work in providing support and 'anticipatory guidance' to address the severe impact

of HD on families. Sands also addressed the collaboration with voluntary organizations such as the Huntington' Society as a source of strength and support for families.

It was clear from these articles that the family was a source of expertise in understanding the individual with HD and in developing supports, but families also benefited from community supports and services designed to augment and strengthen their care-provider role. Yale and Martindale (1984) highlighted the multiple psychosocial issues that individuals and families face, and advocated for a collaborative team approach utilizing social work involvement from the time of diagnosis.

The mainstream literature on Huntington's disease reflects what can be understood as dominant discourses. I will first address these dominant discourses about HD, in order to place my research in the current context of understanding about the illness. I will then highlight the social meanings and implications of HD, an area which noted previously, has been under-researched yet strongly identified as an area of need by families.

Dominant discourses about HD

The academic literature on Huntington's disease is predominantly focused on the trajectory of the illness from a medical perspective and clinical research on finding a cure. The positivist focus places expertise in the hands of those undertaking what is seen as good scientific research – in this area, biomedical researchers. Snowden (2001) suggests that research must also be directed at improving the behavioural aspects of the disease, which often create the greatest challenge for care providers (Snowden, 2001).

Baron (1985) proposes that medical breakthroughs are not accomplishments in and of themselves and only derive significance from what they mean for human beings, their impact on human suffering and capability. The embodiment of the illness, how individuals live their lives with HD, and the day to day reality of their experiences as well as their care-providers encompass the realm of care and caring practices. The capacity to reflect means that humans suffer not only from disease but also from their experience of illness and the cultural meanings that they and others such as family care-providers attach to it (Freund, McGuire & Podhurst, 1991). The over-riding emphasis on finding a cure for Huntington's has meant that little emphasis has been placed on caring for those with the illness (Chiu, 1989a, 1989b). This perspective originally highlighted by the extensive work of Chiu and others in Australia has not resulted in significant changes in caring practices for those with HD in North America and other parts of the world. (Miller, 1976; Smith, 2003; Snowden, 2001; Hardt, 2001; Goolkasian, 2001; Moskowitz, Marder, 2001; Dawson et al., 2004; Aubeeluck, Buchanan, 2006; Kristjanson, Aoun, Yates, Grove, 2006).

Kitwood (1997) in his work with dementia, suggests that the medical model, which is the standard paradigm even when found inadequate or incorrect can maintain certain views or positions by subtle redefinitions, by discounting certain pieces of evidence and highlighting others, or even by suppressing or marginalizing other points of view.

The Discourse of Individual Responsibility

The predominant discourse around health in the Canadian health care context promotes the individual as responsible for his or her own health. Self-care includes the wide range of activities that individuals engage in to promote health, and limit illness and disability (Levin & Idler, 1983, as cited in Penning, 2002). These activities include lifestyle choices a person has made whether healthy or unhealthy and is reinforced by health promotion programs. The responsibility of the individual for his or her own health is also underpinned by cultural values, which view individuals' care as the responsibility of family and community. This emphasis is fuelled by the perception that large bureaucratic structures are now providing care that was traditionally the responsibility of the individual, family and community. The discourse of individual responsibility has particular implications for people with HD, as discussed below.

The social meanings and implications of HD

Identity Loss

Social identity is undermined by chronic illness. Our sense of who we are and our important social relationships are intimately connected with our bodies and their routine functioning. The illness experience has far-reaching social, emotional, moral and spiritual implications (Freund et al, 1991). Nijhof (2002) elaborates on the shame experienced when breaking specific kinds of rules, such as everyday situations, eating and speaking. Although Nijhof's study involved individuals with Parkinson's disease, he extended his

study to include others who are chronically ill, especially those whose symptoms or lack of control may be exhibited in public.

The study of Alzheimer's disease has produced a body of knowledge that addresses identity loss (Orona, 1990). This knowledge is especially helpful in understanding the impact of identity loss in a degenerative illness such as Huntington's disease, even though insight and awareness are thought to be present into the end-stage of HD (Snowden, 2001; Chiu, 1989; Pollard et al. 1999). Identity loss has been described as a dyadic relationship (Orona, 1990) since it profoundly impacts the family as well as the person with HD. For the person with HD the losses invoked by the illness are cumulative, loss of employment, loss of mobility, loss of speech, loss of social relationships. The difficulty in maintaining a sense of oneself, and a sense of coherence, is severely limited by communication difficulties and the attitude and response of care-providers.

The disjuncture from the body

I have linked the body and emotional and cognitive changes together to emphasize the impact of the disease to the whole person. As Henderson (1989) points out in commenting on holistic care, that it is fallacy to consider the mind and body as separate entities (as cited in Benner & Wrubel, 1989). Frank (1991) proposes that when one is well and performing fluidly the body is experienced as oneself. However, when the body is experiencing breakdown, as in chronic illness, the body is experienced as an uncontrollable dense other with a will of its own (as cited in Freund, McGuire, Podhurst,

1991). This disjuncture from the body so clearly described by individuals with HD, also affects family members who intimately witness the changes. The development and maintenance of self is related to the quality of interpersonal relationships, and the significance of experiencing positive regard (Singer, 2001, Buber, 1970, as cited in Surr, 2006). It follows that loss of self may occur if others, care-providers, family members accord a person no recognition and their relationship allows them only to receive and not to give (Laing, 1969, as cited in Surr, 2006).

It has been suggested that doctors of Western medicine have difficulty dealing with the illness experience and view disease as located in the body separate from the individual (Freund et al., 1991). In this model disease is something to be understood by experts. The patient is thus reduced to a passive informant (Bury, 1991, as cited in Freund et al., 1991). This loss of agency and control over one's choices compounds the loss of social identity experienced by the person with HD.

With Huntington's, the body looks and acts in a way that makes the individual stand out from able-bodied others. The body moves in a way that is seen as not normal, with unusual, dancing, jerky movements. People with HD have talked about others seeing them as drunk, or acting afraid of them. The person with HD often slurs their words and eventually loses the ability to speak, therefore communication is limited with others

increasing the likelihood of being misunderstood. Wittgenstein (1922) the philosopher stated, "The limits of my language mean the limits of my world." For someone with Huntington's their world is limited not just by communication but by what can be understood as an inability of the body to act the way it did before. As noted previously,

this disjuncture from the body is heightened as well by the knowledge that a person with HD experiences insight and awareness through to the end stage of the illness, though these abilities are often not recognized by others (Snowden, 2001).

Skirton & Glendinning (1997) in a UK study found that 20% of family care-providers for someone with HD suffered from a stress-related illness. In this study the stresses for care-providers were compounded by the inadequate coordination of care between families and professionals, a lack of awareness of the extent of needs by professional carers, inadequate service provision, and lack of availability of resources.

It appears that historically to present, HD carries a negative connotation or stigma, which adds to the individual and family's struggle in coping with the progression of the illness and finding supportive and informed care.

The struggles faced by people with HD and their families, is especially complicated because of the discourse of individual responsibility. As noted earlier, people with HD often have great difficulty switching attention from one task to another. When the person is held responsible for this, he or she may be perceived as unresponsive or uncooperative. The intellectual and mood changes in HD are likely to contribute to how a person behaves, since how a person behaves reflects the way that she or he thinks and feels (Snowden, 2001). Yet often the person with HD is assumed to be responsible for 'difficult' behaviour, despite behaviour changes being a facet of the illness (Snowden, 2001; Chiu, 1989b). The damage to the brain of the person with HD, which affects mood and impulse control requires an understanding and accommodating response on the part

of care-providers in order to avert or minimize emotional outbursts. The assignment of responsibility and a 'behavioural' label to people with HD mitigates this understanding.

The current study

The study that I undertook responds to some of the shortcomings in current literature and practice regarding HD. The existing literature focuses on the lack of services for people with HD. This thesis takes up that focus from a phenomenological perspective, examining how the lack of appropriate services affects the meaning of the illness and people's lived/existential experiences of illness and caregiving. The meaning of the illness as experienced by individuals with HD and families has received little attention. Yet these understandings can contribute to caring practices that better meet the needs of those with the illness, and to our understandings of illness and care more broadly. This approach also validates the expertise of individuals with HD and families (Benner, 1994).

METHODOLOGY

Research Theory and Design

The influence of the biomedical model has determined what is considered to be good scientific research in the area of Huntington's disease. The positivist approach with its stance on neutrality and objectivity dominates the literature. In the hierarchy of the medical model the doctor is seen as the most influential in treatment decisions. The positivist approach views the input of individuals and families to be biased and therefore less valid. Baron (1985) p.609, a physician proposes, that "a great gulf exists between the way we think about disease as physicians and the way we experience it as people." It has been raised that a "crippling flaw" of the biomedical model is that it does not include the patient and his or her attributes as a person, a human being (Engel, 1978 as cited in McCarthy & Schafermeyer, 2007).

In my study I used qualitative methodology since my purpose was to explore the meaning of the experience of having Huntington's disease for individuals with HD and family care providers. In contrast to the usual positivist approaches, my study acknowledged the expertise of individuals with HD and family care-providers. There has been little research in the literature from these perspectives. This has contributed to misconceptions about the illness, its impact on individuals and families, and a lack of understanding and response in terms of treatment (Snowden, 2001; Hardt, 2001; Chiu, 1989b; Ball, 1982).

My research was informed by interpretive phenomenology, since my research area of interest is about the meanings individuals attribute to their experience of care. This approach pays close attention to interpreting that experience as intimately as possible. I undertook interviews with individuals with Huntington's disease and family care providers to explore fully and in depth their interpretation of their experience with HD in their day-to-day lives, their unique lived experience of health and illness (Benner, 1994) as well as their concerns for their future. The understanding realized through interpretive phenomenology considers historical change, transformations, gains, losses, temporality, and context (Benner, 1994). These understandings provide a way to explore the meaning of HD from individual and family perspectives, and a means of making that experience clearer to others. Benner's is a particular approach to phenomenology that focuses more on social context than is often the case with this approach to research.

My approach to this research links well with approaches necessary for good clinical practice. In my research, understanding is based on knowledge about the illness and perhaps more importantly, empathy and compassion. In studies measuring the qualities that make nurses effective it was found that techniques and knowledge are not enough, and that a caring involvement with a person is required for expert human practice (Dreyfus & Dreyfus, 1986 as cited in Benner & Wrubel, 1989). Of course in social work we are well aware that relationships and an understanding of social context are essential for practice.

Benner (1994) proposes that how the interview is interpreted should fully represent the world of the participants as closely as possible. The phenomenological

approach informed the research process by offering a particular practice of knowledge. Knowledge can be further understood by questioning how some knowledge is considered true and other knowledge is marginalized. We come to see things in particular ways through the concepts and theories we have developed about them. It follows that understanding is imposing a view upon reality. Foucault (1972) suggests that the concepts we develop are a result of “historical” ways of knowing (as cited in Chambon, Irving, and Epstein, 1999). Benner (1989) proposes that the present health care system is designed and controlled based upon treatments and procedures that can easily be counted and priced. Caring practices, healing relationships, and attentiveness that prevent illness and complications and ease suffering are not easily counted or measured and thus become marginalized as the focus intensifies on treatment techniques and procedures (Benner, 1994).

The nature of discourse and whose voices are heard and whose voices are marginalized inform the experience of people with HD and family care-providers. The data derived from the interviews have informed my study and my understanding (Morse & Richards, 2004). As Benner suggests, interpretive phenomenology offers an alternative to quantitative social science studies because it is concerned with individuals’ life worlds and human concerns, habits, skills practices, experiential learning and notions of the good that inform health care practices of help seeking and receiving (Benner, 1994)

Participant Recruitment

Letters of Information were provided to the Huntington Society of Hamilton and Toronto, and staff then passed them on to individuals during scheduled visits or at family support meetings (see Appendix A-Letter of Information). A Question Guide which provided 8 sample questions was attached to the Letter of Information (see Appendix B-Question Guide). The letter detailed information that was relevant to the individual's decision to take part in the study and included the following areas: the purpose in undertaking this study, what to expect as a participant during the study, potential harms, risks or discomforts and the potential benefits of participation. The participants were also informed about their right to withdraw from the study at any time.

It was important for individuals and family care-providers to know what was expected of them in participating in this study. This information is respectful of any participants' time and need to weigh out their decision whether to take part in the study. I am also aware through my knowledge of Huntington's disease that certainty, knowing what to expect, is critical to a person with HD (Snowden, 2001; Pollard et.al., 1999).

Participants

I interviewed six participants, two individuals with HD and four family care providers. Participants included women and men representing a range of ages and stages of Huntington's disease, as well as living and care arrangements. The two participants with Huntington's disease were both approximately 60 years old, one a married female diagnosed with HD about 15 years ago who has been living in a long term care facility

for one year, and a male recently diagnosed with HD who lives with his wife. I also interviewed separately, both spouses of the participants with HD. I interviewed two other family care-providers, an 80 year old male whose wife died of HD two years ago, and a 55 year old woman who has lost several family members to HD.

Ethics

The nature of Huntington's disease and care-giving meant that I had to take specific steps to conduct this research ethically and respectfully. Since the intent of this study is to understand as fully as possible the meaning of Huntington's disease, the lived experience of individuals with the illness and family care providers it was essential that the participants felt comfortable talking about their experiences.

The vulnerability of individuals with HD is heightened by their dependency on family, physicians, specialists, and paid caregivers for aspects of their care. This dependency impedes their ability to speak out about their concerns and views. The need for assurance of privacy was critical and needed to be revisited at different times to ensure a consistent comfort level. Without protecting these rights, participants may have felt threatened that their comments might result in a withdrawal of services, care or support.

Respecting anonymity and confidentiality was particularly relevant since I recruited through the Huntington Disease Society and participants sometimes depend solely on these services for individual and family support. The Huntington Society was not informed of who decided to participate in the study. To further ensure participants'

privacy, individuals with HD and family care-providers if in the same family were interviewed separately. I was aware that individuals with HD may feel inhibited in talking about their experience if it meant expressing criticism of the family member or care-provider they were dependent on. It was important to offer as well a choice of location to ensure the process was accessible and convenient for participants.

This issue of confidentiality was reviewed at the beginning and completion of the interview to ensure participants were comfortable and informed.

The emotional vulnerability of people with Huntington's and the recognition of the severity of the stress on families required an alertness and sensitivity to the impact of the interview process. I discuss this further below.

Interviews

Before I began each interview, I reviewed the consent form with each participant (Appendix C – Consent Form) and offered the opportunity to ask questions. Once the individual's questions and concerns were addressed I asked him or her to sign the consent form. I reinforced at the beginning and before completion of the interview the participant's right to decline any questions they were uncomfortable with or withdraw from the study at any time without any repercussions.

Participants were asked to provide a personal account or narrative of their experience as someone with HD or as a family care-provider. The interviews followed a natural flow from the interview guide, with probing questions when necessary to encourage more in-depth discussion. The intent was to encourage participants to tell their

story of Huntington's disease in a way that was meaningful to them and as they understood it. This was in keeping with the phenomenological approach, which seeks an insider's account and values the expertise of the participant to confirm or negate the researcher's understandings (Benner, 1994). I was also aware of the power differential between the participant and myself as the researcher and wanted to address this imbalance by taking a non-threatening respectful approach that encouraged open and full dialogue.

Since individuals with Huntington's disease experience significant changes in emotion and cognition, it is important to be attentive to these changes during the interview. The knowledge that up to one quarter of people with HD attempt suicide was a strong reminder of the need to sensitize the interview process (Shannon, 2006). My experience in working with individuals with HD and families has provided me with some insight and understanding as to when to pursue a sensitive area of discussion, thereby minimizing or avoiding emotional distress. When interviewing someone with Huntington's disease it is important to recognize the changes caused by the illness. There is often a delay after asking someone with HD a question. It is important to wait and give the person time to respond. It is also easier for the person to be given lots of prompts in conversation rather than be asked a broad open-ended question such as 'What has your experience with Huntington's disease been like?' The person with HD is often distracted easily by noise and commotion around them. It is helpful to be in a quiet room with no other distractions where he or she can fully attend to the questions. The person with HD often has difficulty putting thoughts together and understanding complex information. It

is important to repeat important information and ensure the person understands what has been discussed. It is often helpful to gauge how a person is feeling on a particular day, and if he or she appears to be having difficulty understanding or responding it may be necessary to approach a topic on another day. The person with HD may have impaired reading and writing skills, and difficulty paying attention, reasoning, problem solving and organizing thoughts. An awareness of all of these factors ensures that the person communicating with someone with HD is attentive to the needs of the person and can respond in the most appropriate way. It is also critical to remember that an inability to communicate does not mean an inability to understand. The person with HD has insight and awareness up until the end stage of the illness. Family members are often stressed from care giving and talking about their experiences with HD can be emotionally intensive. These concerns were central in influencing both the pace and direction of the interview. One of the participants with HD seemed to be avoiding a discussion, which was leading into his fears for the future. Rather than pursue that direction and the risk that he was becoming emotionally upset I changed the direction of the conversation. He then appeared more relaxed and shared some of his current experiences. In order to ensure participants felt emotionally grounded and not upset by the end of the interview we returned to talking about the study. We discussed the potential benefits for themselves and their contribution to the academic literature from their participation.

Participants were provided with community contact information to respond to potential difficulties or crises and to ensure they were aware that their emotional response

was respected and validated. Each participant was informed how to contact me if they wished to receive a copy of the completed research document.

Data Analysis

As the audio-taped interviews were transcribed I began analyzing the data, looking for themes. This involved extensive reading, rereading, reflection, writing and rewriting to be able to transform the participants' lived experience into a textual expression of its essence (Morse & Richards, 2004). Using Benner's phenomenological approach to analyzing interview texts (2002) I considered temporality, how one projects oneself into the future and understands oneself from the past. Benner (1994) also subscribes to the notion that reality is socially constructed, people see and understand experiences such as illness through the common linguistic and cultural meanings that they attribute to them. I studied each person's concerns, including what they noticed or identified about their situation, and what mattered to the person. I then focused on all of the texts to look for common meanings (Benner, 1994 as cited in Sinding, Gray, Fitch & Greenberg, 2002). While analyzing the texts I attended to words or phrases that described particular aspects of the participants' lived experiences and reflected on these. Through the process of describing these experiences, I was able to gain more insight into the lived meaning of participants (Morse & Richards, 2002). I also drew on the experiential descriptions from the writings of individuals with HD, and from other literature that explored illness meanings since these layered a deeper understanding (Benner, 1994).

My professional experience working with individuals with Huntington's disease as well as my volunteer role with the Huntington Society has shaped and I believe strengthened my analysis of the transcripts. In particular my relationships with people with Huntington's disease have allowed me some insight and a very detailed, intimate practical knowledge informed by their experiences. As an example, eating becomes a difficult task for individuals with HD due to choreic movements and increasing difficulty swallowing. Sarah who was living in a Boarding home was experiencing difficulty manoeuvring her spoon to her mouth, which slowed down the pace at which she could eat. Despite losing over 30 lbs over a few months, she was told that she "needed to eat faster, that if she couldn't keep up she would need to find another place to live." There was no allowances or flexibility to accommodate Sarah's need for longer time at meals. Thus the provision of care becomes a source of tension rather than support for the person with HD. Knowing people with Huntington's disease helped me to know what to listen for in other people's stories.

My knowledge of these experiences also required that I assume a self-reflective stance throughout the research process to ensure I was not reading into the text what was not there, and projecting my own world into the text (Benner, 1994). Good interpretation is guided by an ethic of understanding and responsiveness to those whose worlds we are interpreting (Benner, 1994). The ethos embedded in existential phenomenology encompasses respect for the social and cultural nature of being human (Benner, 1994).

FINDINGS

The following themes emerged from the analysis of participants' narratives: the challenge to social identity; efforts to sustain social identity; and the mismatch between the needs of people with HD and the available formal care.

The Challenge to Social Identity

As discussed previously, the person with HD loses the ability to walk, talk, think and reason. As these losses accumulate, they ultimately threaten the loss of self. Although the loss of self is attributed to the progression of the illness as in dementia, it is heightened by the response of others. The perception and reaction of others towards the individual with HD is an especially poignant experience for family members and is often accompanied by the withdrawal of friends and families' support. Sally described watching her sister when she was still living at home, walk to the fruit market. "It took so much effort for her that I don't think she noticed the way people were staring at her." Sally strongly identified with the hurt that she felt her sister would have experienced had she been aware of how people perceived her.

Tracy, Neil's wife, talked about the dramatic changes in Neil's life because of how the HD has affected him.

I was thinking my goodness, a piece of his life slowly is being taken away....his job, soccer, he had to give up soccer. He played old timer soccer and he loved it...then the car.

Tracy painfully recounted the changes that Neil was experiencing that were limiting his life as well as her own. Tracy poignantly described how the more symptomatic Neil

becomes, and the less normal he appears to others is hurtfully accompanied by a withdrawal of friends and support at a time when she feels lonely and more isolation. What may be described as the loss of control over one's body is then compounded by the lack of understanding from others.

Sandra, who has HD was always active and enjoyed physical activities such as water skiing , hiking and spending time at her family cottage. Although the memory of those experiences she cherishes, she is now in a nursing home, and as a result of the Huntington's she experiences difficulties with her balance and coordination and requires a walker. She said: " You see other people doing what you can't." It is clear that Sandra's social identity, the way she sees herself has been undermined by the things she can no longer do.

Neil, who has HD has been unable to work or drive for the past year. He is experiencing increasing unsteadiness while walking and is somewhat hesitant to go out alone. When asked about what he like to do with his time, Neil talked about no longer being able to participate in sports; he played hockey and soccer an activity he enjoyed for 25 years. "I mean I like things, but I can't do things." Neil later commented: "I just want... to live a normal life and if I get to the point where I'm going to be put on life support, I really don't want it."

When a person is no longer occupied, boredom, apathy and futility find root. Kitwood (1997) proposes being occupied means to be involved in the process of life in a way that is personally significant and which draws on a person's abilities and powers. It

is obvious that being involved in life and engaged with others is necessary to maintaining social identity. I will expand this point in the discussion section of my thesis.

Efforts to sustain social identity

The physical, communication and behavioural challenges imposed by Huntington's disease have particular social and cultural meanings. As a result, family care-providers become extremely sensitive to the social interactions and experiences of their spouse or family member. It was apparent in my study that family members were striving to sustain the person with HD as a valued social being in a context where they were often viewed as 'out of synch' or 'abnormal'. Family members' attentive responses can be understood along three themes: maintaining social contacts, maintaining social status, and maintaining valued roles. Individuals with HD also strongly identified the need to continue feeling valued and involved in life, and had their own ways of working to achieve this.

Maintaining social contacts

Family care-providers identified the loss of social support for their spouse or family member with HD as well as themselves as one of the most difficult aspects of living with Huntington's. Although the demands of providing round the clock care and supervision to the person with HD limited their contact with others the loss they expressed was compounded by the gradual withdrawal of support from friends and sometimes family. This loss was felt acutely for the person with HD whom they wanted

to protect from hurt. Sean talked about the tremendous effort he expends in order to ensure Sandra has visitors when he goes away since her friends no longer visit on their own volition. Sean stated:

You don't want to have to be calling to arrange it or paying for it, you want it to be friends, who want to be with her. Our friends don't show up, the only way that I can get her friends in to help is that I have to phone them to get them to come in, but there is nobody that just sort of comes all the time and you know that's the sad part. I make a visitor's schedule and I find that friends always come in if they have committed to a schedule.

Sean has also protected Sandra from this information and she believes that her friends continue to be spontaneously involved in her life.

With Huntington's disease, the body looks and acts in ways that makes the individual stand out from others. The choreic movements associated with HD result in unusual dancing, jerky movements. People with HD often slur their words, increasing the likelihood of being misunderstood. The perception and reaction of others towards the individual with HD is an especially poignant experience for family members

Tracy described the isolation both she and her husband Neil have faced as the symptoms of his illness have become more pronounced. She thought if friends or family would come by, even to take Neil for a drive or to the mall it would improve his quality of life. "You know friends don't want to talk to you because they can't understand you or they don't understand the situation." Tracy said neighbours, who were good friends, seemed to react to Neil differently once he had more difficulty speaking clearly, and staggered when he walked. She said that she "hoped Neil didn't notice the way they looked at him."

Tracy recently organized for her and Neil to move to a townhouse complex, which she felt was safer for Neil and more stimulating since many of their neighbours are retired. She hoped he would have the opportunity to make friends. She came to this decision after Neil had fallen near their previous home and no one came to help him.

Maintaining Social Status

Huntington's disease is not only the losses which ravage the body as a result of the illness but about the loss of the person's social status and ultimately one's place in the world. Family care-providers had an intimate awareness of these losses and attempted to maintain the social status of their loved one right through to the end stage of their illness. Considerable research on the connection between social status and health indicates that the degree of control people have over life circumstances, especially stressful situations, and their discretion to act are key influences of health (Public Health Agency of Canada).

Ken's spouse Donna who was diagnosed with Huntington's in her 60's had grown up in a large family in Saskatchewan. Her father had died of Huntington's, which resulted in financial hardship for the family and necessitated Donna leaving school early. As a result she was proud of her independence and her accomplishments. Ken described the importance that she attached to having a driver's license as being

kind of a status symbol for her. I went to her doctor and asked him not to take her driver's license because she really prides that and she wasn't driving anymore. He never did take her license away, she had it right up to the end ... it was an idea that she treasured.

A driver's license also represents freedom and adulthood, cultural meanings that Ken was also sensitive to. He was able to take comfort in knowing that Donna was able to keep her

license, which was an achievement she valued and an important connection to the social standing she once had.

Sally spoke about her mother's experience, having lost several family members to Huntington's disease. Sally's sister Sharon had just been admitted to a chronic care facility and Sally's mother visited her every day, and socialized with the other patients as well. Reflecting on her mother's relationships with Sharon and other patients Sally commented:

They enjoyed being with her and my Mom enjoyed being there and she said that was her life now. She had arthritis, but going into the care facility to spend time with her daughter and other people gave her a reason to get up and going now.

Sally felt that her Mother's relationship with Sharon and other patients with HD was reciprocal and enriched herself as well as the patients. The development and maintenance of self is related to recognition and equality in interpersonal relationships (Singer & Buber as cited in Surr, 2005). It follows that loss of self and status may occur if care-providers and family accord a person no recognition and their relationships only allow them to receive and not give. As Sally observed with her sister and others:

You can see in someone with Huntington's that they are still the person, especially if you have grown up with them, you know that they are still inside, that the body is reacting in this awful crazy way, but you know that when they smile or laugh they are getting it, but they can't express it with words.

Sally, like her Mother, was able to continue to engage with her sister and other patients with HD, in a way that conveyed they were part of a reciprocal relationship not just passively receiving care.

Sandra was well aware that many people saw her only in the context of her illness and in a sense her social identity, who she was as a person before the illness was no

longer recognized. In our interview, Sandra wanted to talk about her contributions as a teacher and the importance she places on being remembered as a person beyond her Huntington's diagnosis. To help others learn more about her, she would like to write her life story and share experiences she had as a teacher. "As a teacher I had so many things...I kept a letter by a student I taught in the 70's and she went on to become a doctor." Sandra took pride in the appreciation the student had shown her and wanted others to know the contribution she has made.

Maintaining valued roles

Family care-providers were painfully aware of the losses experienced by their loved one with HD and strove to maintain the roles that the individual valued in his or her life. The loss of these roles, were vivid reminders of the life they had before the illness.

Ken talked about his wife Donna when she was no longer able to look after the homemaking tasks that she used to as being a particularly difficult time for her. Ken said:

At that point I sat down with her, and we talked about things and she gave me instructions on how to wash clothes, how she wanted things done... various cooking instructions she wanted followed, so I jotted this all down in a little notebook.

As Ken assumed more of the household jobs, he would then seek approval to see if what he was doing was right. He said he did that, "So that she was still a part of things, she was running the show even when she wasn't physically doing it, but she was still under control." Essentially Ken was sustaining Donna in the role of homemaker by his careful attention to the tasks she used to perform and his turning to her for confirmation

that the role was being enacted well. Maintaining valued social roles helped to provide a sense of meaning to the person with HD's life and a sense of purpose.

Kitwood (1997) proposes that to be a person, is to live in a world where meanings are shared, thus the nature of our interactions are made meaningful in a cultural setting.

Mismatch between the needs of people with HD and available formal care

Participants in this study described several instances in which the needs of the person with HD were not met – or worse – in formal care settings. In some cases this 'mismatch' seemed to be the result of health professionals' lack of knowledge about the illness. In other cases it was clear that the behaviours of a person with Huntington's disease, which disrupt 'standards' of time and action, were not accommodated at all – and this failure of care was made worse by current conditions in the health system. In other cases inadequate care or abuse seemed to emerge from health professionals blaming people with HD for their behaviour.

Ken talked about the difficulty in finding a family doctor with experience in HD and getting emergency treatment for Donna. "The thing that disappointed me most was the unawareness of the various emergency room staff. They have not been exposed to Huntington's, they don't know what it is." Ken expressed his frustration "cause you are coming to them for help and you expect them to at least know what she is going through and know something about it and they knew nothing about it." Ken described one occasion when Donna had fallen and dislocated her shoulder:

So I took her to hospital and they gave her a sling and I said, 'it is not much use, you either strap her arm down or don't bother because by the time I get her home that sling will be gone,' and by the time I got her home it was.

Ken's difficulties in finding informed medical care for Donna added to his heightened sense of responsibility. He described his dilemma, feeling that he needed to be the one to inform medical staff – staff to whom he had hoped to turn for help about the illness.

For family care-providers, one of the most disruptive changes has been described as a profound slowing down of the individual with HD in almost every area that affects functioning, walking talking, thinking and reasoning. This slowing down impedes the person's ability to maintain a 'usual' pace of action – yet the kinds of care that are offered (both home care and institutional care) require the ability to keep up. Sean talked about the time Sandra requires to get ready to go somewhere.

The person slows down, the person slows down immensely; it's not 10 seconds, it's maybe 10 minutes to an hour. You have to realize if the doctor says she wants us there at 10:30am, I need to be at Sandra's at 9:00am, since it takes 15 or 20 minutes to get her ready and get her down to the car and then you go to the hospital and then it takes another 15 or 20 minutes to get to the doctor. You got to build all these time parameters in.

It was clear however, that most formal care systems did not build those time parameters in – far from it. Ken, whose wife Donna died two years previously had been a full-time caregiver for ten years. He shared that at one point he had looked into several nursing homes for her.

By this time I was feeding her, so I said to one nursing home, 'How about meal times, what happens, and she says, 'she will have to feed herself.' I said 'well she can't feed herself,' she said, 'well she is only allowed 15 minutes if they feed her' and I said 'it takes me an hour to feed her.' So I thought if I need to come in and feed her, I may as well keep her at home.

Individuals with Huntington's disease often end up in psychiatric institutions

even though they do not have a mental illness, in nursing homes even though they are not elderly, and labeled as behaviour problems even though certain behaviours are a facet of the illness. Many people with Huntington's while comparatively young end up in long-term care facilities despite the ill fit of these facilities to meet their needs (Dawson, Kristjanson, Toye & Flett, 2004; Kristjanson, Aoun, Yates, 2006; Moskowitz, Marder, 2001; Hardt, 2001; Smith, 2002). People with HD have the same needs for social stimulation as people without HD yet others around them may not realize this due to their communication difficulties, such as delayed or limited speech. People with HD also require continuity in their care, knowing what to expect each day and a clear understanding of their social schedule (Pollard et al., 1999). In the nursing home where Sandra lives, neither consistent social activities nor continuity were available.

Sean, Sandra's husband describes what he did to address these shortcomings of the care system. When Sandra was still living at home Sean approached Home Care for assistance. He was able to get assistance one hour per day, five days per week. Although this was helpful to "get Sandra bathed and get her going in the morning" it did not address her need for social stimulation. As a result, Sean hired two companions to spend time with Sandra every day for several hours to occupy her days. He interviewed fourteen potential companions and "chose those whose personalities I thought were compatible, but also very important that the person needs to have empathy."

When Sandra moved to the nursing home, Sean continued to pay for companions to spend time with her there. Although Sandra has slowed down in all the parameters mentioned above, she sees herself as a "young 60 years old" and with different interests

than the older persons in the same care facility. She referred to the support she derives from the companions in helping her cope with the limited social activity and interaction in the nursing home. “I don’t know how I would live. It’s not a stimulating enough environment. They are all 80.”

Sean commented later in the interview when talking about the extended family that he grew up in how he might not have been so isolated trying to care for Sandra. “It was a lot healthier when we had supports. Now we are expecting the community to do the supports and the community doesn’t know you too well.” Aware that having people ‘know you’ and behave empathically along with mental stimulation were important to Sandra, and aware that the care facility could not provide either at a level he saw as necessary, Sean organized them himself. His care and commitment are apparent. Yet as he says himself, the cost of hiring companions to make up for the shortfalls of the system is not within everyone’s financial means.

Blaming people with HD

Huntington’s disease has been found to damage selective parts of the brain, leading to specific difficulties in thinking, which in turn give rise to specific and predictable changes in behaviour (Snowden, 2001;Hardt, 2001; Chiu, 1989b). It has been proposed that individuals with HD should not be held totally ‘culpable.’ The extent to which the person with HD is held responsible varies depending on the care-provider’s understanding of the illness, and may affect the care that the individual receives.

Family care-providers interviewed for this study identified education about HD mostly provided by the Huntington Society support group or newsletter, aided them in their understanding, and shaped how they related to their ill spouse or family member. Sean referred to what he saw previously as Sandra's stubbornness and her inability to see another point of view, both of which created significant tension in their relationship. He then clearly highlighted how Sandra's diagnosis of HD helped him understand why she behaved in certain ways:

That was a turning point in my life because from then on I treated her completely different and we got the counselor involved and I got really involved. I was a much more understanding and compassionate person.

Sean learned that as a result of the Huntington's disease, Sandra was unable to change her focus easily and once upset, had difficulty managing her feelings and response. Sean responded to Sandra differently after her diagnosis by avoiding getting into arguments. This meant that both he and Sandra were less frustrated and Sean was able to circumvent potentially emotionally charged situations. Sean recognized that Sandra was not culpable for her behaviour and was able to make changes in the way he related to her.

It seemed that in situations where health professionals had more time and fewer patients, less blaming occurred. Sean compared the nursing care of Sandra in her private room as opposed to when she shared a ward:

The nurses that were in the section where the private room was were extremely empathetic and showed it and didn't say she was crazy like the other ones did...some of them were not very nice, you know.

Sally's brother's care experiences both in the general hospital and in the psychiatric facility identified profoundly the mismatch between her brother's needs and

the care available – a mismatch that resulted in part from health professionals seeing Jeff as responsible for his behaviour.

In the mid-70's Sally's brother lived at home with their elderly mother. The family, perceiving their mother was overly stressed, decided to take their brother to the closest general hospital. She said that staff called her mother several times complaining that he refused to bathe. Sally said:

They knew nothing about Huntington's....we went up to see him and he was gone, they had shipped him to the psychiatric institution and they didn't tell us...he was right in with the mentally-ill people.

In discussing how staff seemed to approach her brother Sally commented: "I think it was, 'well we need to do this and we need to do it now.' They just didn't know how to handle things." The first time Sally's mother went in to shower him "he had bruises all over his back; he had been beaten."

In contrast to the staff who could not bathe him, and who assaulted him, Sally commented that her mother was able to "do anything with him", which addressed the different way his mother approached him with compassion and understanding, as opposed to force. Family care-providers accounts suggest that a family member's love for the person with HD, and their understanding that he or she was not responsible for difficult behaviour, made anything possible, whereas in the institution the lack of love and understanding was stark.

Sally also talked about the care her brother received at the facility they moved him to, which specialized in providing care to people with chronic illness. "They knew sometimes they could have an outburst...and they didn't see it as weird, they knew it was

part of the disease.” Sally feels her brother’s quality of life was much better since staff seemed to understand the illness and treated him with respect. Similarly, Sally described her sister’s care in a chronic care facility. In the physical caring, the nurses talked with her and learned about her life. In the process of learning about her as a person Sally said: “They liked her there too and one of the nursing staff said that they regarded her as a sister.” Sally felt that the kind of caring that her sister experienced, based on an understanding of her illness, allowed her to feel safe, cared for and loved. Thus the kinds of emotional upsets and behaviours often attributed to HD that are seen as problematic in many care settings did not occur with Sally’s sister. Seeing the individual as responsible for the emotional cognitive changes that are caused by the HD affects the care-provider’s attitude and response. This likely has a significant impact on both the frequency of emotional upsets that the person with HD experiences and the management of those upsets.

DISCUSSION

Within a biomedical model, self-assessments of health are often attributed little value and are not seen as providing valid representations of health (Penning, 2002). However, research evidence has shown that self-reports are often more accurate than physician evaluations for predicting various outcomes, including mortality. Ferraro & Farmer's study (1999, as cited in Penning, 2002). As well, in contrast to the biomedical model, the World Health Organization (WHO) defines health as "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity" (WHO as cited in Penning, 2002). In this more holistic approach, not only their disease defines individuals and quality of life in each of these spheres is a more complete measure of one's health and well-being. As the WHO definition suggests – and this research confirms - individuals with HD and families can make a significant contribution to health care providers regarding their physical, mental and social needs.

This research, in taking a phenomenological approach, considers study participants as experts in their own experience. Study participants drew our attention to key themes: challenges to social identity; sustaining social identity which encompass maintaining social contacts, maintaining social status, and maintaining valued roles; and the mismatch between the needs of the person with HD and the available formal care.

As Cassell (1982) points out, "people suffer from what they have lost of themselves in relation to the world of objects, events and relationships" (as cited in Freund, McGuire & Podhurst, 1991 p.149). Affliction and suffering thus results, not only from the illness, but from the threat to the coherence of a person's world (Freund et al.,

1991). It is apparent in this study that caregivers of people with HD, and people with HD themselves, have to work hard to maintain the coherence of a social world with many losses.

The ultimate goal of caring has been described as the commitment to the protection and enhancement of human dignity. This dignity is embedded in the meaning of the experience for the individual patient (Gadow, 1980; Wros, 1994). The behaviours of a person with Huntington's disease disrupt norms of time and action. Protecting their dignity requires health professionals to make allowances for these behaviours by not blaming the person, and making time to ensure the person is not rushed, prodded or made to feel inadequate for not keeping up. In Miller's (1976) research, as in this study, family members emphasized certain characteristics of good care: empathic, flexible, accessible and responsive to individual and family needs. These findings were substantiated by Yale and Martindale's study (1984), which also concluded that lack of knowledge about the issues families face with HD inhibited good practice and that care provided was often fragmented, offered at a late stage in the illness, or not at all.

Participants in this study talked about the losses of abilities experienced by the person with HD and particularly the changes to the body. It is clear how HD departs from certain kinds of societal norms especially norms dealing with how bodies behave, and norms about time. Family members were sensitive to the amount of time it takes the person with HD to perform any activity such as getting dressed or eating. Family members were able to accommodate these changes in the person by allowing more time,

and maintaining a flexible attitude and schedule. In care facilities this kind of flexibility is not readily accommodated and the person is expected to keep up.

Behaviour changes as well as changes in mood were identified by families as one of the most difficult challenges in caring for the person with HD. However, in Miller's study and mine, when families were informed how Huntington's disease influenced the behaviour of their loved one, it helped them understand and manage these changes, and develop strategies to minimize emotional outbursts. This does not seem to be the case in many care facilities. In some instances individuals with HD were treated with frustration and annoyance and seen as responsible for their behaviour. It is likely that the care-providers attitudes and frustration contribute to the emotional outbursts experienced by the person with HD. It may be, that these behaviours that are seen as problematic and disruptive by many care-providers and facilities could be averted or avoided by an informed compassionate approach.

It is important to emphasize that the kinds of care available to people with HD are affected by wider social policies and dominant discourses. The central positioning of the biomedical model and the medicalization of health and issues related to health care have a significant far-reaching impact on the care and treatment of individuals with chronic illness such as Huntington's disease. The discourse of the biomedical model as proposed by Foucault (1972, cited in Chambon, Irving, and Epstein, 1999) is accorded more value or truth than other discourses, particularly those of the patient or family. Individuals with Huntington's and family members struggle to have their needs met in a system where their needs are marginalized.

In a care facility or institutional setting, responding to individuals with HD is limited by a myriad of factors including the individual care-provider's attitude and knowledge of HD, lack of educational opportunities such as in-services, and the staff to patient ratio which limits the time available to spend with assigned patients. Since institutional care is a provincial responsibility there is variation across the country and no policy to regulate accommodation, funding arrangement, or even quality and standards of care (Tarman, as cited in Penning, 2002). In Ontario, Bill 140, known as the Long Term Care Act has no set standard for the number of hours of daily care that each resident in a long-term care facility receives, and staffing ratios average one front-line worker for twelve or more residents (Moir Welsh, Toronto Star, as cited in Hamilton Spectator, July 30, 2007). The person with Huntington's disease often requires assistance with activities of daily living, such as dressing, bathing, eating. Family members who took part in this study talked about the slowing down of the person in all these areas and the amount of time needed to perform daily tasks. This is also compounded by the difficulty that persons with HD have in initiating any activity. It is clear that if a care provider has twelve or more residents to assist, that the person with HD will be hurried and unable to keep up, or left unattended for extended periods. This coupled with the lack of knowledge of HD means that the person with HD is misunderstood, may be treated with frustration, and may also be labelled uncooperative or non-compliant.

In recent years the types of care that are provided to people both at home and in institutions have been eroded by fiscal restraints. Thinner service provision places pressure on professional care-providers to pare down their services (Aronson, 2002;

Aronson & Sammon, 2000). In social service work it has been noted that, staff cuts coupled with standardized work create pressured environments where work is speeded up, which then places restrictions on the types of skills, tasks and relationships that workers can form with clients and supervisors (Aronson & Sammon, 2000). In institutions that provide highly routinized, standardized care no allowances are made for people who can't do things the way everyone else does. The fast pace of care requires that people have to keep up. The ability of health professionals even to feed someone at a pace that makes sense for them is also less and less possible. In this study, we see that the kind of care and empathy that comes with knowing a person, the ability to take the time to move at the person's pace, and the mental stimulation that people with HD require are rarely available.

It appears that in care facilities the emphasis placed on standardized caring routines rather than personalized approaches, results in sporadic rather than consistent caring practices. The opportunity to learn ways of responding to someone with Huntington's falls to individual care-provider's interest or motivation rather than being accepted practice. As Benner (1989) proposes, as society places more value on technological advances, skillful, compassionate care that is less measureable, becomes marginalized. Family members in this study strongly identified the need for informed compassionate care and viewed this as more important than other aspects such as care-provider qualifications. When this type of care was provided they were able to identify a change in the mood and overall quality of life for the person with HD and a reduction in their own stress.

In Miller's (1976) research as in this study, individuals with HD valued the consistent support and understanding of a counselor to see them through the course of the illness. In Miller's research counseling was made available to individuals with HD and families; in my own study, this was sporadically provided due to limited funding to Huntington's Societies for individual and family support. Families felt that counselors outside of the Huntington Society would be uninformed about HD, thus individuals with HD and families often went without the support that they determined would be helpful.

Continuity of care has been defined as how individual patients experience integration and coordination of services (Haggerty, Reid, Freeman, Starfield, Adair, & McKendry, 2003). The care of a person over time is also seen as an intrinsic part of continuity and is especially relevant for those with chronic illness such as Huntington's disease. Family members in this study highlighted their difficulties in finding informed care, a lack of consistent information, and feelings of uncertainty and insecurity as a result. Persons with HD also pointed out an uncertainty around their care provision and a sense of isolation rather than support. Yale and Martindale's study (1984) addressed the failure of professionals to communicate adequately and the lack of a collaborative approach as barriers to comprehensive care, which impeded rather than augmented the families' ability to cope. It has been established that when care is long term as in the care of someone with Huntington's disease consistency and flexibility are critical. This flexibility is necessary to adapt to the changes in the individual's needs over time.

Although as noted, family members provide up to 80% of the long term care for individuals with HD, this care is often provided in isolation and without adequate support

for both the care-provider and the person with HD. Care-providers experience significant stress and loss of quality of life, along with the loss of quality of life for the person with HD. It is apparent that individuals with HD are placed in nursing homes since there are few alternative care facilities and family members are significantly stressed and no longer able to provide the level of care necessary at home. Home health care services were designed with the intent of potentially limiting the numbers of people requiring long-term residential care, yet as the families in my study indicated there was little support available to assist them in the care of their relative with HD. Although the intent of home health care is to provide a range of supportive and therapeutic services to help persons who are incapacitated to live at home the non-medical supportive services provided such as meals-on-wheels, homemakers and friendly visiting are viewed as secondary. These non-medical supportive services tend to be ideologically placed within the realm of family or friends and are not funded by national health insurance (Chappell, as cited in Penning, 2002). The acute care needs such as medical, nursing and related services are given priority. Families with financial resources can access privately paid community and residential care. Those without resources or family are most likely to end up in nursing homes, homes for the aged or other institutional facilities due to the limited availability of publicly funded community based care (Penning, 2002). People with Huntington's disease are often unable to work when the disease begins to manifest, often in the person's mid-thirties or forties resulting in financial hardship for families which then precludes their ability to access private home care services. It is apparent that the lack of

publicly funded community based care directly impacts on the lives and choices of individuals with Huntington's disease and their families.

In considering the mismatch between the needs of the person with HD and care facilities, perhaps the most glaring misfit is in placing a relatively young person in their 40's or 50's in a nursing home where other residents are in their 80's. The losses accumulated by someone with HD, coupled with the expectation of being placed in a nursing home for their final care permeates the despair that often informs the illness. Yale and Martindale's study (1984) confirmed these findings and discussed the dilemma for families when faced with the inappropriate placement of some people with HD to geriatric or psychiatric wards, despite the fact that the person is neither old nor mentally ill. As noted previously the person with Huntington's disease has the same need for social stimulation as those without HD, yet the ability to communicate and initiate contact and activities with others is often impeded by the illness. Social activities and interaction with others are often viewed by care facilities as the individual or family's responsibility as well as a non-essential service and are thus affected by the paring down of resources. Families in this study identified the moral dilemma they were faced with knowing that the care their relative with HD received was inadequate, yet there was limited community support such as home care to augment the significant emotional and physical demands placed on the care-provider in caring for the person with HD at home.

For family care-providers the painful decision to place their family member with HD in a nursing home is compounded by a lack of choice. For the person with HD, the right to self-determination is often compromised by the symptoms of the illness or the

care decisions are assumed formally or informally by others such as care-providers.

Snowden (2001) notes that patients who participate in making decisions are less likely to be behaviourally disruptive than those from whom all choices have been taken.

At the same time it is apparent from this study that individuals with HD in facilities that offered compassionate informed care were able to draw on spiritual reserves necessary to sustain a sense of coherence or meaning in life. Hardt (2001) a person with HD has recommended 'assisted living homes' staffed by knowledgeable professionals to help those who are still capable of living by themselves to maintain their independence. There is only one assisted care facility in Canada for people with HD located in Toronto, Ontario (R. Andrejas, personal communication, May 1, 2007). Hardt (2001) also focuses on the need for expertise, training and funding to establish HD-friendly nursing homes, staffed by HD-competent professionals which would provide further choices based on suitability. This care ideally involves the collaboration of many disciplines over the trajectory of the illness. These include physiotherapists, occupational therapists, recreational therapists, nursing, social work, physicians and specialists, such as neurologists.

Family members in my study were extremely sensitive to how their family member with HD was treated by care-providers. Family members valued empathic people who recognized the person with HD was not culpable for his behaviour. Families saw this as critical above all else in ensuring the person with HD was treated with respect and dignity. As noted previously, techniques and knowledge are not enough and will not

work unless a basic level of attachment and caring are in place (Dreyfus & Dreyfus, 1986; Henderson 1989 as cited in Benner & Wrubel, 1989).

In referring back to the social science literature that related to this study several important areas were identified that confirmed my own findings: the value of family support and expertise, the need for comprehensive flexible care provision including community care, the need for continuity in overall care provision, and the need for residential care facilities that specialize in the treatment of those with HD. In Miller's work (1976) as in my own study, family members were key in defining the kinds of services that were needed

It is important to note that there was little social science literature related to the meaning of the experience of Huntington's disease for those living it. Helder and colleagues (2002) raised this concern in their study and concluded that systematic research on the psychosocial aspects of HD is scarce. As a result there appears to be limited awareness of the devastating psychosocial impact of the disease on individuals with HD and family members (Klimek et al., 1997 as cited in Dawson et al., 2004; Snowden, 2001). Although the social science literature search on HD did not provide current research, the research discussed in this study addressed the impact of the illness on individuals and families and also addressed the inadequacies of the community wide response. These findings continue to highlight that the emphasis on advances in research in the search for a cure for HD have not been paralleled with an emphasis on the tremendous social impact of the disease for individuals with HD and families. This lack of attention has contributed to a lack of understanding of the disease, inadequate care and

supports available to buffer the ravages of the illness, and a sense of isolation and despair for individuals and families. As Miller's study advocated 30 years previously engaging individuals with HD and families in our understanding of the illness and in the creation of adequate supports and services, may be a way to move families from the margins to the centre and begin to address the long-standing social injustice linked to the disease.

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Appendix A
Letter of Information



Title of Research Project: Huntington's disease: Time to Care

Student Investigator: Marnie Wilson
Graduate Student: Master's in Social Work

Student Faculty Supervisor: Christina Sinding,
Associate Professor
Department of Social Work
McMaster University
Hamilton, Ontario, Canada
(905) 525-9140 ext. 22740

Why are we doing this study?

I would like to know what you think as someone with Huntington's disease or as a family member about the care that you receive now. I am interested in hearing about the supports and services you have. I would also like to know what ideas you have on services and supports you feel would improve the quality of care you receive, what is helpful and what is not helpful. I am also interested in hearing from you as someone with Huntington's disease or as a family care provider what supports you feel would improve the quality of care that you receive.

I would like to understand from your perspective what the gaps are in the supports and services you receive and advocate for the changes that you feel would be helpful. I want to know about your experiences with the services that you receive. I would like to talk with you even if you are not receiving services at this point since your contribution about what you feel would be helpful in your care is a valuable part of this study.

What will happen during the study?

You will be asked to participate in one interview, at a time convenient to you for approximately 60 minutes in length with myself. The interview will be tape recorded with your permission in order to accurately represent your thoughts and ideas. The interview will be in a location of your choosing and convenience.

During the interview which will be in the form of a conversation, I will be asking you about your experience with Huntington's disease, the kind of supports you have now, as a person with Huntington's disease or as a family care provider, what services and supports you feel are needed and your ideas on what supports you feel would improve your quality of life. I will also ask you for some demographic information like your age and living environment.

If you decide before, during or after the interview that you no longer wish to continue or take part in the study, it is your right to withdraw at any time.

If you have Huntington's disease and are living with a family care provider who is also a participant in this study I will meet with you separately and in private to ensure you have the opportunity to talk about your experience. If necessary, I may contact you again by phone after our interview to clarify information that we talked about.

Potential Harms, Risks or Discomforts:

I am aware that in talking about your experience with Huntington's disease you may experience strong feelings or become upset. Please remember that you do not need to answer any questions that make you feel uncomfortable or that you do not want to answer.

As a family care provider you may also feel upset when talking about your experiences in providing care or attempting to arrange care for a loved one. You do not need to answer any questions that make you feel uncomfortable or that you do not want to answer. If you choose not to continue with the interview it is your right to withdraw at any time. If you become upset and decide that you may need further support or assistance I have provided contact information for community services that will be able to provide support.

Potential Benefits

I would like you to have the opportunity as a person with Huntington's disease for your voice to be heard and acknowledgement that your experiences are valued.

As a family care provider I would also value your contribution and the opportunity to hear your thoughts and ideas regarding care needs. I hope from talking with both people with Huntington's disease and also family care providers it will provide a better understanding about the social care needs of people with HD. Your contributions will help fill a gap in the research literature on Huntington's disease. At present there is little research available from the perspective of individuals with Huntington's and families.

I would like to use this research to advocate for community resources for the supports and services you have identified.

Confidentiality:

Anything that we talk about in our interview, both individuals with Huntington's disease and also family care providers, that could identify you will not be published or told to anyone else, unless I have your permission. Your privacy will be respected. I will not be sharing your name or identifying information with anyone at the Huntington Society or with any other person, family member, or agency. If you are living in a Boarding home or long term care facility your information will not be shared with any staff at the facility. Your identity will be kept anonymous. If you are living with a family member and are participating in the study I will interview you at a separate time at your convenience to ensure your privacy. Your information will not be shared with your family care provider or any other service or agency. The information we talk about and the tape recording of our interview will be kept in a locked filing cabinet in my home office. It will be kept for three years and then will be destroyed.

What if I change my mind about participating in the study?

Your participation in this study, and your decision to talk with me is voluntary. It is your choice to be a part of this study or not. If you decide to participate, you can decide to stop at any time, even after signing the consent form or part way through the study. If you decide not to participate there will be no consequences to you. No one will be informed of your decision not to participate. If you decide to withdraw at any point, any information that we talked about including our tape recording will be destroyed unless you have indicated otherwise. If you do not want to answer any of the questions, you do not have to, but you can still be in the study. Your decision whether or not to participate will not affect any services or supports you are currently receiving.

Information About the Study Results:

You are welcome to contact me if you wish to receive the study results. The research findings will be made available to you and a copy of the research document will be provided if you wish.

Information about being a Research Participant

If you have any questions or would like more information about the study itself, please contact myself, Marnie Wilson, (905) 521-0696 or Christina Sinding, my Research Advisor at McMaster University School of Social Work, (905) 525-9140 X22740

This study has been reviewed and approved by the McMaster Research Ethics Board. If you have concerns or questions about your rights as a participant or about the way the study is conducted, you may contact:

McMaster Research Ethics Board Secretariat
Telephone: (905) 525-9140 ext. 23142
c/o Office of Research Services
E-mail: ethicsoffice@mcmaster.ca

Consent for Contact

If you think you may be interested in participating in this study, or would like to talk with me about the study before making your decision, I would like to ask your permission to contact you by phone or email. Your decision whether to participate in the study or not will not be shared with any staff at the Huntington Society unless you decide to do so. In that way your privacy and anonymity will be respected. I give permission for Marnie Wilson to contact me regarding my possible involvement in her research study. She may contact me by phone/email at _____.
The best times to reach me are at _____.

Additional Comments (if any)

Name: _____

Date: _____

Appendix B
Question Guide

In my research I am going to be exploring the social care needs of individuals with Huntington's disease and family care providers from each of their perspectives.

It will be a qualitative research study, using semi-structured research questions, which will generate discussion about individuals' experiences with the supports and services they are receiving, and their ideas about the kind of care that would improve their quality of life. I will be asking the following questions of both people with Huntington's disease and family care providers.

Interview Guide:

1. I am thinking of social care as the kind of supports and services that are available to assist you with Huntington's disease. Can you tell me about the supports and services you have now?
2. What has your experience been with finding care?
3. What are the gaps in the care that you receive?
4. Are there any services that you feel would make life easier for you?
5. What kind of supports do you feel would improve your quality of life?
6. What do you feel is important in your care?
7. What is your biggest worry about your care?
8. If you could make a change in the care you receive now, what would that be?

Appendix C
Consent form



Student Investigator: Marnie Wilson

Student Faculty Supervisor: Christina Sinding
Associate Professor
Department of Social Work
McMaster University
Hamilton, Ontario, Canada
(905) 525-9140 X22740

Why am I doing this study?

I would like to know what you think as someone with Huntington's disease and as a family member who cares for someone with Huntington's disease about the care that you receive now. I am interested in any supports and services that you have. I would also like to know what ideas you have on services and supports you feel would improve the quality of care you receive. I am interested also as a family care provider what supports and services may be beneficial to you.

I would like to hear your thoughts on what the gaps are in the care you receive and advocate for the changes that you feel would be helpful. I am interested in your experiences with these services. I would like to talk with you even if you are not receiving any support or services at this point as a person with Huntington's or a family care provider, since your contributions about what you feel would be helpful in your care is a valuable part of this study.

What will happen during the study?

You will be asked to participate in one interview with myself, at a time convenient to you, approximately sixty minutes in length. The interview will be tape recorded with your permission in order to accurately represent your thoughts and ideas. The interview will be in a location of your choosing and convenience.

During the interview which will be in the form of a conversation, I will be asking you about your experience with Huntington's disease, the kind of supports you have now as a person with Huntington's disease and as a family care provider, what services and supports you feel are needed and your ideas on what supports you feel would improve your quality of life. I will also ask you for some demographic information, such as age and living environment.

Potential Harms, Risks or Discomforts:

I am aware that in talking about your experiences with Huntington's disease you may have strong feelings or become upset.

Please remember that you do not need to answer any questions that make you feel uncomfortable or that you do not want to answer.

If you decide that before, during or after the interview that you no longer wish to continue or take part in the study it is your right to withdraw at any time.

Potential Benefits

I hope that we can understand more about the social care needs of people with Huntington's disease from the perspective of people with Huntington's and family members who are caring for someone with Huntington's disease.

I would like you to have the opportunity for your voice to be heard and acknowledgement that your experiences are valued.

I feel your contribution as a person with Huntington's disease and as a family care provider will help fill a gap in the research literature on Huntington's. At present there is little research available from the perspective of individuals with Huntington's and families.

I would like to use this research to advocate with community providers for the supports you have identified.

Confidentiality:

Anything that we talk about in our interview that could identify you will not be published or told to anyone else, unless I have your permission. Your privacy will be respected. I will not be sharing your name or identifying information with anyone at the Huntington's Society or with family members. Your identity will be kept anonymous. If you as a person with Huntington's disease and one of your family members are both participants in this study I will arrange the interviews at different times to ensure your privacy and confidentiality.

The information obtained by me, and the tape recording of our interview will be kept in a locked filing cabinet in my home office. It will be kept for three years and then will be destroyed.

What if I change my mind about participating in the study?

Your participation in this study is voluntary. It is your choice to be part of the study or not. If you decide to participate, you can decide to stop at any time, even after signing the consent form or part way through the study. If you decide not to participate there will be no consequences to you. If you decide to withdraw at any point, any data you have provided to that point will be destroyed unless you indicated otherwise. If you do not want to answer some of the questions you do not have to, but you can still be in the study. Your decision whether or not to participate will not affect any services or supports you are currently receiving.

Information About the Study Results:

Please contact me if you wish to receive study results. The research findings will be available to you and a copy of the research document will be provided if you wish.

Information about being a Research Participant

If you have questions or require more information about the study itself, please contact myself, Marnie Wilson, at (905) 521-0696 or Christina Sinding, my Research advisor at McMaster University School of Social Work, (905) 525-9140 X22740

This study has been reviewed and approved by the McMaster Research Ethics Board. If you have concerns or questions about your rights as a participant or about the way the study is conducted, you may contact:

McMaster Research Ethics Board Secretariat
Telephone: (905) 525-9140 ext. 23142
c/o Office of Research Services
E-mail: ethicsoffice@mcmaster.ca

CONSENT

I have read the information presented in the information letter about a study being conducted by Marnie Wilson of McMaster University School of Social Work. I have had the opportunity to ask questions about my involvement in this study, and to receive any additional details I wanted to know about the study. I understand that I may withdraw at any time, if I choose to do so, and I have agreed to participate in this study. I have been given a copy of this form. I have also been informed that the interview will be tape-recorded if I am in agreement. If I am not in agreement the interview will not be tape-recorded.

I am in agreement with the interview being tape-recorded. Yes No

Name of Participant

In my opinion, the person who has signed above is agreeing to participate in this study voluntarily, and understands the nature of the study and the consequences of participation in it.

Signature of Researcher or Witness

