HUNTINGTON DISEASE: DEATH, PSYCHIATRY AND HEALTH SERVICES

by

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ABSTRACT

Huntington Disease (HD) is a genetic, degenerative neurological illness that affects mood, cognition and motor movement. Complications from the disease result in death 10 to 15 years post-diagnosis. However, individuals may be aware that they have HD years before the development of the most severe effects. Drawing on in-depth, qualitative interviews with 20 individuals with HD and 10 informal caregivers, this thesis explores the salient experiences that occur outside of predictive genetic testing. Findings emerged in four areas; namely, attitudes towards death and suicide, experiences with psychiatry, interactions with health services and bureaucracies, and the needs and experiences of caregivers. Participant accounts also form the basis for a number of health services recommendations, including trajectory-specific support groups and the need for disability benefits that are sensitive to the realities of HD. The thesis also explores the numerous ambiguities that HD creates for conceptual categories, specifically in relation to diagnosis, death and illness experience.
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INTRODUCTION
The experiences of the ill have been the source of numerous social science investigations (e.g., Briggs & Mantini-Briggs, 2003; Frank, 1991; 1995; 2004), which provide an interpretation of sickness grounded in the accounts of individuals who are ill, in contrast to biomedical explanations of disease. However, such accounts of illness experience have primarily focused on infectious diseases (e.g., Briggs & Mantini-Briggs, 2003; Farmer, 2001) or the advanced stages (i.e., hospitalization) of chronic conditions (e.g., Frank, 1991; Bluebond-Langner, 1996). In particular, the experiences of individuals with diseases of comparably longer duration and chronicity are absent from this literature.

This absence is resonant for three reasons. First, within industrialized nations, chronic diseases and lasting disability are prominent health concerns. For instance, Seale (2000) has noted that, while industrialized nations have substantially increased life expectancy due to better treatment of infectious diseases, the age at which disability begins has not changed over time. Accordingly, although individuals gain additional decades on their lifespan, chronic illness and disability are likely to be present throughout these years. Second, as the accuracy of medical technology increases, individuals will have a better understanding of their genetic susceptibility to disease (Davidson, MacIntyre & Smith, 2008). Thus, people can become aware of risks to their health well before hospitalization and often before the most severe symptoms are experienced. Third, accounts of illness experience have improved our ability to understand, treat and communicate about disease (e.g., Frank, 1991; 1995; Boyd, Johnson & Moffat, 2008; Kleinman, 1978). As such, these overlooked areas of disease experiences have received few of the benefits of social science health research.
Given these omissions, this thesis focuses on the experience of Huntington Disease\(^1\) (HD). The disease is both fatal and degenerative, meaning that individuals deal with an increasing number of disabilities and symptoms as it progresses to its terminal stage (Paulsen, 2004). Furthermore, since 1993, a genetic test has existed for HD, allowing individuals who are at-risk for the disease to determine whether or not they have inherited the mutation decades in advance of typical symptom onset (MacDonald, 1993).

Drawing on qualitative interviews with twenty individuals with HD and ten informal caregivers, this thesis aims to document the illness experience of HD. This investigation will focus on four main areas. First, given the long duration of the disease, as well as the ability to learn of one’s disease status well before serious symptom onset, this thesis will explore how individuals with HD conceptualize death and dying.

Second, HD also has a number of psychiatric symptoms and is often characterized by specialists as a “neuropsychiatric disease” (Benjamin et al., 1994). However, there is both a long history and an extensive number of psychiatric case studies detailing the psychiatric misdiagnosis of HD (e.g., Appollonio et al., 1997; Duesterhus et al., 2004; Jardri et al., 2007). As such, this thesis’ second area of focus will be experiences with psychiatric care, symptoms and particularly the psychiatric misdiagnosis of HD.

Third, as the disease has diverse effects (psychiatric, cognitive and motor), individuals with HD encounter a number of care providers, such as HD clinics and general practitioners, and health bureaucracies, including providers of disability benefits. Following previous health research (e.g., Boyd, Johnson & Moffat, 2008; Bungay, 2008), the third main focus of this thesis will be providing participant based recommendations for improved health services.

Fourth, although the disease physiologically affects individuals, non-diagnosed family members also face a number of HD-related consequences. It has been argued that informal

\(^1\) The physiological aspects of the disease are detailed in the next section.
caregivers, such as spouses and parents, deliver more care services than medical professionals (Lowit & van Teijlingen, 2005), and have been found to experience substantial economic and psychological stress as a result (Carretero et al., 2009; Dunkin & Anderson-Hanley, 1998; Williams et al., 2009). For these reasons, tied with its genetic nature, HD has been referred to as a “family disease” (Brouwer-DudokdeWit et al., 2004; Sobel & Cowan, 2001). Accordingly, another area of investigation will be the accounts of these informal caregivers, both in terms of their experiences as caregivers and their perspectives on the previously mentioned HD-related areas.

Lastly, after presenting findings that speak to these four topics, this thesis advances an argument on the problems that HD poses for categories, both within medicine and medical sociology. Specifically, the disease inserts ambiguities into conceptualizations of death, the locus of disease, and division of medical knowledge. Moreover, rather than refining categories, or creating new ones, this work argues that diseases such as HD can be understood by including these ambiguities and by creating meaningful contrasts between categories.

These areas of investigations address a number of gaps within the HD literature. In particular, they highlight the experiences of the HD community outside of genetic testing, whilst forwarding a number of health services recommendations. This thesis also provides an example of illness experience research that differs from dominant research trends. Specifically, this thesis provides accounts of non-hospitalized individuals dealing with a chronic, disabling and terminal condition. Although illness perspectives such as these are not currently well represented within research, as life spans and technology improve, they are increasingly likely to be representative of illness conditions in industrialized countries.
An Overview of Huntington Disease

Huntington Disease is a hereditary genetic condition that impacts movement, affect and cognition (Paulsen, 2004). The disease is autosomal dominant, meaning that if one parent has the mutation, each child has a 50% chance of inheritance. The illness usually affects adults between the ages of 30 and 45. However, symptoms can also appear in children and young adults; when this occurs, the illness is classified as Juvenile Huntington Disease (JHD) (Paulsen, 2004). Approximately 1 in every 10,000 individuals has HD (Huntington Disease Society of America, 2009; Huntington Society of Canada, 2009). Although a rare condition epidemiologically speaking, HD affects an estimated one in every 1,000 Canadians through family, friendship and professional networks (Huntington Society of Canada, 2009).

The effects of the disease are caused by the accumulation of a protein (the huntingtin protein) within neurons. As the body ages, it becomes increasingly susceptible to the effects of this protein and the correlated neural cell death (Huntington Society of Canada, 2009). As neurons continue to die, the brain compensates by sending stronger neural signals. These signals result in the awkward and jerky muscle responses characteristic of HD. Individuals’ ability to move, eat, and communicate with others is subsequently compromised. The death of neural cells also causes cognitive deterioration, including reduced thought processing abilities, memory recall and decision-making skills (Quarrel, 1999). The cognitive effects of the disease may also result in psychological side effects, such as increases in anxiety, depression, and blunted affect (Bates, Harper & Jones, 2002). Individuals may experience difficulties in perceiving emotional changes, both within themselves and others. The disease is also connected with increases in impulsivity, which may result in the development of compulsive pathologies, such as excessive drinking, drug use or gambling (Bates et al., 2002). Complications from HD symptoms (e.g., pneumonia, malnutrition) result in death, which typically occurs 15 to 20 years after diagnosis (Huntington Society of Canada, 2009). Although HD cannot be treated, its psychological
consequences may be addressed through the use of psycho-pharmaceuticals and/or therapeutic
counseling sessions (Bates et al., 2002).

The production of the mutated protein that causes the aforementioned symptoms is
controlled by trinucleotide cytosine-adenine-guanine (CAG) repeats. While everyone has CAG
repeats, individuals with repeats of 40 or higher will develop HD in the course of a normal
lifespan (Rosenblatt, Ranen, Nance & Paulsen, 1999). Additionally, those who have repeats
ranging from 36 to 39 may also experience symptoms of the disease, though not until later in life
and with decreased severity (Langbehn et al., 2004). Accordingly, one method of HD detection is
through a genetic test assessing an individual’s number of CAG repeats (Quarrel, 1999).

Individuals over the age of majority with a family history of HD may undergo genetic testing to
determine whether they will develop HD prior to experiencing any symptom onset.

While predictive testing can inform someone if they have the gene for HD, it cannot
determine whether they have already begun to express HD symptoms. The results of the genetic
test are thereby not considered diagnostic of HD (Rosenblatt et al., 1999). Instead, a formal
diagnosis of HD is made through a series of motor and cognitive exams, which indicate the
presence of the initial stages of the disease and actual neural damage. However, as noted above,
HD also has a number of psychiatric symptoms, and while the disease is diagnosed based on a
cognitive-motor exam, growing evidence suggests that individuals may experience psychiatric
and cognitive effects well in advance of the characteristic motor symptoms (e.g., Duff, Paulsen,
Beglinger, Langbehn & Stout, 2007).

The disease also has implications for those who are not directly physiologically affected.
Resources distributed by the Huntington Society of Canada (HSC) routinely state that HD is a
“family disease”. That is, while the disease is physiologically located in an individual, a
diagnosis of HD has implications for the family as whole (e.g., Brouwer-DudokdeWit et al.,
2004). These implications include decisions about communicating genetic risk to offspring,
making reproductive choices, and caring for individuals with the disease (Cox & McKellin, 1999; Decruyenaere et al, 2004, 2007). Individuals within HD families that receive negative test results may also experience “survivor’s guilt” (Meiser & Dunn, 2000; Tibben, 2005).

**A Brief History of Huntington Disease**

Early accounts of chorea date back to Ancient Egypt, and connect the loss of motor control to brain dysfunction (Hayden, 1981). During the Middle Ages, HD was called St. Vitus’ Dance; this term was used to both classify individuals with a type of kinetic mania and describe those with movements similar to HD, long before the disease was differentiated from other motor movement disorders (Wexler, 2008). Indeed, it was not until the 15th century that Paracelsus connected the disease to the central nervous system (Hayden, 1981).

During the 19th century, the disease received attention from one of the most seminal figures in neurology, Jean-Martin Charcot. Contrary to then-contemporary medical opinion, Charcot (1887/1987) demonstrated that neurological diseases, such as HD and ALS, were discrete ailments, rather than different manifestations of the same phenomenon. The disease, however, would be most systematically described by its namesake, George Huntington. Huntington was a third generation doctor in a town that had numerous families affected by HD. (Wexler, 2008). Drawing on his experience, and that of his predecessors, Huntington was the first to discover the hereditary quality of the disease, a particularly impressively feat given that Mendalian findings on heredity yet to be popularly disseminated within medicine (DeJong, 1953). Huntington presented his findings in his article *On Chorea* (1967/1872), a work that led Sir William Osler, perhaps one of the most influential doctors of the modern age, to state that “in the history of medicine, there are few instances in which a disease has been more accurately, more graphically or more briefly described” (Durbach & Hayden, 1993).
After Huntington’s description, HD became both famously and infamously associated with genetics. During the early 20th century, the disease became the target of eugenics movements within the United States. For instance, Davenport and Muncey (1916), after completing a survey of HD families, called for the forced sterilization of individuals with the disease. Canada had similar policies, as evidenced by Alberta’s 1942 legislation amendment permitting the sterilization of individuals with HD (Cairney, 1996).

Recently, the disease has had a more positive association with genetics. In 1983, an American-Venezuelan research cohort discovered the approximate location of the HD gene (Gusella et al., 1983), and in 1993, as referenced above, the precise genetic location of the disease was discovered (MacDonald, 1993). This discovery marked the first time that an inherited disease had been genetically located. These advances allowed the creation of the aforementioned predictive genetic test for HD that was also the first of its kind (Gusella et al., 1983; MacDonald, 1993).

The disease also underwent a change in terminology in the late 20th century. Early accounts of the disease focused on its kinetic nature; the disease was termed “dancing mania” or “St. Vitus’ Dance” in the middle ages, and the term “chorea”, the actual phraseology used by Huntington, is drawn from a Greek term for dancing. However, as discussed above, the disease contains numerous other features, and some individuals may experience comparatively minor chorea (Rosenblatt et al., 1999). Accordingly, in 1972 the term “Huntington(s) Disease” began to replace “Huntington’s Chorea” (Wexler, 2008). While “Huntington’s Chorea” is still used within academic literature, the illness appears more accurately described through this change in terminology, a change that has been adopted by both of the major North American HD societies (Huntington Disease Society of America, 2009; Huntington Society of Canada, 2009).
Social Science Research on Huntington Disease

A substantial amount of research has been conducted on the physiological and biomedical aspects of HD (e.g., Gusella et al., 1983; Huntington, 1967/1872; Macdonald, 1993; Rosenblatt et al., 2004), which has played an important role in improving the lives of individuals with HD. However, as Frank (1997) suggests, benefits can also be derived by presenting perspectives other than the biomedical, particularly the perspectives of the individuals who actually embody the disease. Indeed, Wexler (2008) has noted that the majority of professionals focusing on HD are clinicians, while others (e.g., Gibson, Timlin, Curran & Wattis, 2004; Nygaard, 2006; Roger, 2007) have stated that current research has not done an adequate job of involving individuals with the illness.

That said, there have been several investigations into the experiences of individuals with HD, with Alice Wexler’s two books perhaps being the best known. The first, *Mapping Fate* (1996), provides a memoir of growing up within an HD family, as well as chronicling Alice and her sister Nancy’s involvement in HD research and the development of the predictive test for the disease. Wexler’s second work, *The Woman Who Walked into the Sea* (2008), is perhaps the most comprehensive sociological work on the disease to date. Wexler traces the history of HD within America from three different vantage points: that of the HD community, the medical community, and the connection of HD to eugenics. Wexler describes the diversity of experience of individuals with HD and highlights their treatment’s variability across communities. She also connects HD to larger movements within science, particular genetics/eugenics, and discusses how this affects our current perceptions of the disease.

Likewise, a Vancouver-based researcher, Sue Cox, has also contributed numerous important works on HD. In particular, Cox’s work focuses on experiences around predictive testing (1999; 2002; 2003; Cox & McKellin, 1999), a topic that remains resonant within social science research. These studies were the first to outline the many dilemmas that unfold for HD
families around the issue of predictive genetic testing, such as how family members communicate about genetic risk (Cox & McKellin, 1999). Cox has also discussed how individuals at-risk of developing HD request a genetic test and compiled numerous accounts of individuals’ test-taking experiences for the HSC (2002; 2003).

In many ways, the majority of HD research within the social sciences is a continuation of Cox’s work, such that the preponderance of research focuses on predictive genetic testing. Several reports have focused on issues leading up to the test. For instance, a recent report described how individuals seeking predictive testing utilized “microdecisions” to expand their perception of time leading up to the procedure (Scully, Porz and Rehmann-Sutter, 2007). Similarly, Taylor (2004) observed that individuals considering taking the genetic test situate their decision within a broad nexus of issues (e.g., how the test may affect their social relations) and also defended their right “not to know” by refraining from testing. In a second study, Taylor (2005) stated that decision-making vis-à-vis predictive testing was more complex than a test/no test dichotomy, noting that decisions instead varied on two axes (openness/non-openness and engagement/non-engagement). Similarly, Forrest and colleagues (2003) noted that families felt a sense of obligation and responsibility to communicate genetic risk to younger generations. In a later study (Forrest et al., 2005), the team discovered several nuances around family disclosure of genetic status, including the gendering of risk disclosure and subjective accounts of who was considered “family”. In their most recent study (Forrest et al., 2009), young people reported several discrete strategies that their families used to inform them about HD, including having always known about the disease, being told gradually, and keeping HD a secret.

Recent research has also detailed the consequences of genetic testing. For instance, Decruyenaere (2002) has noted that individuals who receive a positive test have more negative feelings and are avoidant of issues relating to HD. Decruyenaere and colleagues (2004) also described changes in marital relations after the genetic test, stating that although the gene-
positive cohort had higher reported marital satisfaction than their partners, the majority of individuals (70%) remained married. In a later study (Decruyenaere et al., 2007), reproductive decision-making were analyzed amongst a gene-positive cohort, noting that the majority (58%) decided to have children, but utilized embryonic diagnostic procedures. Researchers have also outlined issues relating to genetic discrimination that have resulted from the creation of the predictive test (Bombard et al., 2007; 2008), noting that the availability of predictive testing can have negative implications for an individual’s employment status, insurability and ability to access health care resources. They also documented several strategies employed to cope with potential genetic discrimination (keeping low, minimizing, pre-empting and confronting), which varied according to actual experience with discrimination (Bombard et al., 2008).

While predictive testing is by far the most popular subject within social science literature on HD, a few reports have also focused on the experiences of families living with a diagnosis of HD. Smith et al. (2006) discuss family members’ process of dealing with JHD, with a particular focus on the process of first noticing the disease to dealing with the “relentless process” of JHD. A more recent exploratory and phenomenological analysis of JHD described both the difficulties that family members had in understanding such a unique illness and their associated feelings of isolation (Brewer et al., 2008).

Although there are numerous in-depth investigations into HD, the experiences of individuals who are gene-positive or with disease onset are almost completely absent within the literature. While both Cox (1999; 2002; 2003; Cox & McKellin, 1999) and Bombard (2007; 2008) discuss the experiences of individuals who are gene-positive, their primary focus is on issues relating to genetic testing or their new genetic status. Novas’ (2003) study of HD web forums and chat rooms included both symptomatic and at-risk individuals, but similarly focused on issues of genetic risk. In one of the rare reports not focusing on genetics, Dawson et al. (2004) interviewed individuals with HD (and caregivers) around their needs for palliative care, and
found that individuals struggled to find both pertinent information and secure healthcare resources, as well as reporting fearing for the future. Roger (2007) also focused on end-of-life issues, with particular emphasis on memory loss. Like Dawson (2004), she noted that individuals with HD had an apprehension about the future, while also trying to adapt to their new disabilities and changing social relations.

While biomedical studies regularly include individuals diagnosed with HD, comparatively few social science studies include symptomatic and gene-positive individuals within their samples. Instead, as seen above, the focus is predominately on “at-risk” individuals or the loved ones/caregivers of symptomatic individuals. Several critiques of HD research over the past several years have highlighted both the predominately biomedical accounts of the disease (Wexler, 2008) and the absence of individuals with HD in research (Gibson, et al., 2004; Nygaard, 2006; Roger, 2007). Accordingly, given both the brevity of the literature and these recent critiques of HD research trends, this thesis aims to explore the many salient issues for both caregivers and individuals living with HD, aside from their already well-documented experiences with predictive testing and genetics.

**Project Overview**

Based on information presented above, this thesis aims to fill a gap in the HD literature by addressing the yet-unexplored issues outside of genetic testing. This gap is particularly important given the unique trajectory of HD, where individuals receive a diagnosis of a fatal disease years in advance of the most debilitating effects. Additionally, as both studies on genetic issues and the disease societies highlight, HD has implications for the entire family (e.g., Brouwer-DudokdeWit et al., 2004; Huntington Society of Canada, 2009). Consequently, this disease is arguably best understood by incorporating numerous perspectives, rather than just those of the sufferers alone.

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2 See Appendix 1 for a complete discussion of methodology.
Accordingly, this project will describe the experiences of individuals living with HD, as well as those of their informal caregivers, with an emphasis on salient issues outside of genetic testing. In addressing this gap, this thesis also aims to provide a number of the benefits that social science research has previously delivered, such as feedback on medical services, and suggestions for improved client-doctor communication (e.g., Boyd, Johnson & Moffat, 2008; Frank 1995).

**Sample**

Twenty individuals (see Table 1 for summary of demographics) with the HD gene mutation were recruited for this study, with all but three having previously received the neurological diagnosis confirming symptom onset. The physiological effects of HD on the participants ranged extensively, from the three aforementioned asymptomatic participants, to two other participants currently residing under managed care. Participants’ ages ranged from 23 to 83 (M = 54); 12 of the 20 participants were male.

<table>
<thead>
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<th>Table 1. Demographics</th>
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<tr>
<td><strong>People with HD</strong></td>
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Ten caregivers were also recruited for the study. Nine of the caregivers were the partners of individuals with HD (eight wives and one husband), and one was a grandmother. Several caregivers attended to multiple individuals. For instance, one participant was caring for her husband, son and granddaughter. The ages of the caregivers ranged from 37 to 64 (M = 54). While the majority of individuals with HD in this sample had partners, not all partners volunteered to be interviewed. This cohort is therefore composed of 7 individuals in dyads and 3 individuals who were interviewed individually.
Recruitment

Participants were recruited across British Columbia with the aid of a social worker operating in a Vancouver Huntington Disease Resource Centre (HDRC) and a second social worker operating on Vancouver Island. Individuals were contacted by these social workers either via e-mail or at their monthly support groups. Interested individuals were given the author’s contact information and a flyer explaining the nature and purpose of the study. Snowball sampling was also used to recruit eligible individuals who were related to interviewees.

Data Collection

Qualitative, in-depth, semi-structured interviews were the primary source of data (Lofland & Lofland, 1995; Weiss, 1994). The majority of the interviews (N = 26) occurred in person and were conducted at a time and place of the participant’s choosing, which, with one exception, was the individual’s personal residence. Due to the distance between their residence and where the research was being conducted, four participants were interviewed over the telephone. All interviews were digitally recorded and ranged from thirty minutes to two and a half hours in duration. With a few exceptions, both members of a dyad were interviewed in the same day, one after the other, with the participants deciding on the sequence. Every interview was conducted in complete privacy. Upon completion of the interview, the author took detailed fieldnotes, including descriptions of the interview location, relevant participant behaviour, and researcher-participant interactions (Emerson, Fretz & Shaw, 1995; Morse & Field, 1995).

Data Analysis

Data analysis was framed by a number of previous works within the fields of philosophy, medical sociology and medical anthropology. Authors such as Nietzsche (1977; 2000),
Kierkegaard (1989), Frankl (1984) and Merleau-Ponty (1962) contributed to the author's perspective, understanding, and interest in investigating individuals' relationship to illness, death and experience. While this thesis does not employ Frank's methodology (1995), his work (1991; 2004) on illness experience was influential in grounding this investigation in the accounts of ill individuals. Anthropologists, such as Klienman (1978), Farmer (2001) and Briggs (2003) also provided important insight.

That said, this thesis, while not a grounded theory project per se, drew on numerous aspects of that methodology, particularly in regards to its previous use in qualitative health research (e.g., Bottorff, Kalaw, Johnson, Stewart, Greaves, & Carey, 2006; Glaser & Strauss, 1964; 1965; 1968; Johnson, Moffat, Bottorff, Shoveller, Fischer, & Haines, 2008). Specifically, data analysis utilized line-by-line analysis, thematic analysis and case comparison. Line-by-line analysis was conducted on each transcript to generate large codes to efficiently organize the data (Strauss & Corbin, 1998). This coding structure was then used to code the data using NVivo qualitative data management software. Coding runs for each major code were generated and were then subsequently read multiple times to develop emergent and inductive themes. Case comparison analysis, utilizing the Microsoft Excel Spreadsheet program, was also conducted to compare each individual case across the entire participant sample (Miles & Huberman, 1994). This technique allowed the data to speak to the particulars of an individual case, whilst also representing aggregate trends. The resultant themes form the basis for the proceeding findings section. Analyses were not guided by any particular theoretical perspective, instead, as suggested by several researchers (e.g., Glaser & Strauss, 1967; Latour, 2005), theories were utilized to advance specific sections of the analysis.

Having reviewed data analysis, data collection, sample details and the relevant literature, the findings of this thesis will now be presented. The findings are collected in four chapters, which will discuss death, psychiatry, care services and caregiver experiences, respectively.
CHAPTER 1: PERSPECTIVES ON DEATH AND DYING

Introduction

Complications resulting from HD typically cause death within 15 to 20 years after diagnosis. A diagnosis can only be made by a neurologist and is only pronounced after the initiation of symptom onset (Paulsen, 2004). However, there is a genetic predictive test available for HD that allows an at-risk individual to know whether they will develop HD within a normal lifespan (Macdonald, 1993). As such, individuals with HD, or those who are gene positive, may learn that they will develop a fatal illness well in advance of the appearance of the major symptoms.

While there have been numerous explorations on death and dying (e.g., Charmaz, 1980; Charmaz, Howarth, & Kellehear, 1997; Lawton, 1998; Seale, 1998, 2000), no study as of yet has considered the perspective of individuals living with HD. Indeed, when previous studies have engaged individuals who are dying, they tend to do so well into the terminal stages of the illness (e.g., Glaser & Strauss, 1965; Lawton, 1998). Individuals living with HD offer a contrasting perspective, as they may be cognitively aware of the terminal nature of their disease well in advance of its physiological effects. This perspective is particularly important given recent research suggesting that future advancement in genetic testing may present individuals with susceptibilities to other conditions with comparable foreknowledge (Davidson, MacIntyre & Smith, 2008). Consequently, the experiences of the HD community may allow us to gain insight on how death and dying are conceptualized by individuals who learn of the risks to their mortality prior to the materialization of these risks.

This chapter engages with these issues by presenting perspectives on death and dying from individuals living with HD. The chapter describes the HD community’s numerous perspectives on death and suggests new avenues of investigation for social research on dying,
particularly as understandings of medicine and health continue to be influenced by advancements in genetic technologies.

**Literature Review**

Social research has often been accused of neglecting both the embodied nature of human beings and their mortality (e.g., Frank, 1995; Hawthorne & Leaman, 2001; Schilling, 1995). In many ways, this critique remains accurate; this can be noted, for instance, in the complete omission of a discussion of death and dying in Rose’s influential work on biomedicine, *The Politics of Life Itself* (2007). However, while arguably a neglected area of research, death and dying has attracted the attention of some of the most important theorists in sociology. Both Durkheim (1897/1952) and Simmel (1918/2007) focused on death and dying, as did later theorists, such as Berger (1967) and Giddens (1991). Of these, Simmel’s essay *The Metaphysics of Death* (2007) stands as a vigorous, if infrequently cited, conceptualization of death and dying. While writers such as Giddens and Berger discuss death as an antagonist to life and security, Simmel sees death and life as co-creating one another. That is, to be a living being is to have the very boundaries of your existence defined by the omnipresence of your eventual and certain death.

While perspectives on death and dying may have been forwarded by some of the most prominent social theorists, empirical research on death and dying was largely neglected until mid-twentieth century (cf. Durkheim, 1952). It was at this time that Faunce and Fautlon (1958) wrote a manuscript calling for more attention to be given to end of life issues, suggesting the need for an empirical “sociology of death and dying”. However, it is Glaser and Strauss’ (1964, 1965, 1968) pivotal works on death and dying that perhaps stand as the most important and defining investigations within the field. Their research took place within hospitals and focused on individuals with terminal illnesses. Most famously, they outlined the numerous “awareness stages” that an individual may arrive at, or pass through, whilst learning of the terminal nature of
their condition (Glaser & Strauss, 1965). They argued that a patient’s ability to access these stages was dependent on the amount of information and type of communication they received from medical professionals. Their findings further suggested that medical professionals were controlling the avenues by which individuals could engage with their mortality, as well as contradicting current medical logic at the time by asserting that patients would be better off with complete information on their health, rather than being spared the negative aspects.

In the spirit of Glaser and Strauss’ work, many recent sociological discussions on death have focused on the secluded sites where dying occurs. More specifically, it is the taboo nature of discourse around death in contemporary Western culture and the actual isolation and segregation of individuals who are dying that has become a source of interest (e.g., Giddens, 1991; Seale, 1998; Walter, 1991). These obscured sites of dying have been the centre of attention of numerous empirical investigations into the dying process. For instance, Lawton’s (1998) influential ethnography, describing the process of dying within care homes and how people were treated as their bodies began to fail them, led her to outline notions of “good dying” in opposition to “dirty dying”.

During the same period that Glaser and Strauss were conducting their influential work, psychologists were developing similar stage based theories that would also have a profound impact on understandings of death. For instance, Parkes (1983, 1996), drawing on Freud’s conceptualization of grief as both a goal-orientated activity and a potential source of pathology, outlined a stage based theory of grief as a natural part of the dying process that must be moved through. Most famous amongst the stage-based theories of grief, death and dying is Kubler-Ross’ (1970) description of the five stages that individuals facing death progress through: denial, anger, bargaining, depression and acceptance. Indeed, Kubler-Ross’ stage theory has almost become ubiquitous with death and dying.
Both the empirical and theoretical work on death and dying within the past several decades has been characterized by stage-based theories, with Glaser and Strauss' (1965) "awareness stages" and Kubler-Ross' (1970) five stages of grief being amongst the most influential. However, in recent years, these stage-based theories have faced numerous critiques regarding their universality and applicability (e.g., Cheung & Hocking, 2004; Valentine, 2006). Specifically, both Timmermanns (1994) and Mamo (1999) have suggested that Glaser and Strauss' conceptualizations emphasize only the cognitive experience of dying, omitting its emotional aspect. Reed and colleagues (1994) suggested that, contrary to "acceptance" being a desirable outcome of individuals coming to terms with their death, it may actually increase unhealthy behaviours (e.g., smoking and substance abuse), therefore decreasing survival time. Likewise, Kingbury (2000), both a clinical psychiatry professor and a family member of an individual with a chronic illness, has questioned the applicability of such theories to the context of long-term illnesses. Patterson (2001) described similar problems with stage-based theories, noting that chronic illness does not follow a direct trajectory, rather oscillating between the foregrounding of both wellness and illness.

The study of death and dying has also arguably inherited Glaser and Strauss' (1965), as well as other prominent contributors' (e.g., Illich, 1975; Sudnow, 1967), focus on medical systems and their subordination of the individual, as well as their concentration on the experiences of those in the acute or terminal stages of their illness (Exley, 2004; Walter, 1993). Thus, the experience of those who are dying, particularly those who are dying over a longer period of time, has been omitted, while the perspectives of institutions, family members and those in acute care (particularly for cancer) has been emphasized (Exley, 2004). Consequently, there appear to be growing critiques of the earlier perspectives and consensus on the numerous limitations within the literature, particularly in the neglect of the experience of chronic illnesses.
Seale (2000) has also suggested that death and dying are already well understood in relation to “degenerative” diseases in Western countries, although it is important to note that “degenerative” in this context predominately refers to cancer and heart conditions. Instead, he argues that increased attention should be given to infectious diseases with long courses, such as tuberculosis and HIV/AIDS. As Exley (2004) notes, this rather limited disease focus is indicative of a larger trend in death and dying research to focus almost exclusively on individuals with cancer and neglect those suffering from non-malignant conditions.

This neglect is certainly the case with HD, as there are no studies of the experience of death and dying in this disease; there is also scant literature that recounts experiences comparable to those of the HD community. Some notable exceptions are Bluebond-Langner’s (1996) and Lawton and Gabe’s (2003) work on cystic fibrosis and Sweeting and Gilhooly’s (1997) article on Alzheimer’s Disease. However, two of these studies focused their interest on the experiences of family members of individuals living with chronic illness. Furthermore, while Lawton and Gabe’s (2003) article did focus on the individuals suffering from cystic fibrosis, the discussion primarily centred on adults living with what is presumed to be a “child’s disease”.

Only Wexler (1996; 2008), Roger (2007), Dawson et al. (2004) and King (2005), in her case study of a child who had passed away from JHD, explicitly discuss issues of death and dying in relation to HD. However, King’s focus, like many other studies of death and dying, is on the medical system and suggestions for treatment, as opposed to the experiences of the individual. In contrast, Dawson’s study (involving nine individuals with HD and 19 informal caregivers) highlights both the fear of the future within the HD community, as well as the need for greater and better organized palliative care. Like Dawson, Roger’s report (five individuals with HD) described anxiety about the future, as well as difficulties dealing with new disabilities and changing social roles. While Wexler’s first work (1996) primarily focuses on her own experiences with her mother’s HD, her latter work (2008) does describe the deaths of several
individuals with HD, particularly Phebe Hedges, who committed suicide by walking into the ocean and drowning herself. Wexler does offer a good discussion of some of issues relating to death and dying, such as stigma and suicide, but her project is primarily historical.

While Wexler describes some historical cases of suicide, self-harm has been connected with HD more intimately since the advent of predictive genetic testing. In their policy paper on testing protocols, Benjamin et al. (1994) discuss the importance of taking a medical and family history that emphasizes suicide attempts. Indeed, suicide rates in individuals with HD may be as much as 12 times higher than that of the general population (Farrar, 1986; Paulsen, 2001). These rates are also higher than individuals with comparable disorders (e.g., brain trauma) (Druss, 2000; Harris, 1997). However, as Paulsen et al. (2005) note, the vast majority of the work on suicide and HD focuses on issues on predictive testing. Their quantitative analysis of individuals both at-risk and diagnosed with HD (N = 4171) suggests that suicide risk fluctuated throughout an individual’s experience with the disease. The period before the neurological exam and the period where the individual began to lose self-sufficiency were main stages in which individuals experienced suicidal ideation.

In summary, aside from reports on suicide rates and risks, issues around death and dying have received little attention within the HD literature. Currently, it is perhaps the resources distributed by HD societies, such as the Huntington Society of Canada (e.g., Paulsen, 2004; Rosenblatt et al., 1999), as well as their respective websites, that contain the best discussion of death. Nevertheless, issues of death and dying only comprise a few pages of these resources, as they are more focused on providing general descriptions of multiple aspects of HD, rather than a detailed analysis of issues involving palliation. Thus, there remains a need for descriptive and experiential accounts of the specific issues involved with death and dying within the context of HD.
On Finality, Chance and Probability

While suicide has attracted a great deal of attention within literature on HD, virtually nothing is known of how people with HD perceive dying from their disease. Interestingly, the majority of participants, despite their diagnosis with a fatal illness, still viewed their lives as open-ended. Indeed, numerous participants stated that they were not distressed about the terminal nature of HD. For instance, Kenneth (76, diagnosed in 2003) repeatedly emphasized that he still intended to live to an older age than both his mother and his father. Accomplishing this task was almost a form of competition for him, which he refused to let his diagnosis affect. He said, “It just gave me all the more reason to say “stuff it guys I’m going to live right though – right on through”.

My dad did – mind you he left at 72 but what the heck. Better diets now so I’ll last till 92 at least”. Kate (57, diagnosed 2005) echoed these statements by noting that she was not afraid or concerned about the ending of her life. This attitude was particularly noteworthy for her, as she had been afraid of death since she was very young as the result of an unfortunate experience at a hospital. However, her experience with HD, as well as comforting her mother and partner as they passed away, helped her adopt a new perspective. She said, “I don’t really have any fears myself now you know, maybe when I’m actually heading off there and it’s all happening I will again a bit you know. And I might have to read some books, but right now I don’t have any fears.”

Though both Kate and Kenneth have been diagnosed with HD for several years, neither of them perceived the disease as truncating their lives.

Critically, this attitude was not “denial” of their condition, as, with just one exception, each participant acknowledged that they had HD and that HD was a fatal disease. Instead, individuals emphasized that their lives were still open-ended. While Kenneth accomplished this through resistance, and Kate through acceptance, the majority of participants used discourses around chance and probability to deemphasize deterministic interpretations of their future and to contextualize death from HD as similar to other forms of death.
The most common avenue by which chance and probability were inserted into discussions on death was the phrase “I could get hit by a bus tomorrow”. Many participants uttered this statement (or a variation of it) during the interviews when the topic of death from HD was raised. As this statement quite efficiently communicates, having a fatal condition, such as HD, does not equate dying from that condition. While this perspective might be considered a type of black humoured optimism, it also effectively removes the finality from an individual’s biography. Furthermore, many participants followed a statement of this type by also noting that the interviewer could similarly be “hit by a bus tomorrow”. So not only is their death indeterminate, but those who are presumed to be “healthy” may also die much earlier than they will. Additionally, as these statements suggest, everyone presumably has an equal chance of being hit by a bus. As such, having HD does not exclude them from dying from another quite common cause or outliving numerous people with no comparable diagnoses. The disease, therefore, could not be used to definitively write the last chapter of their lives.

Indeed, Keith (55, diagnosed in 2003) related the details about one type of accidental death that he felt at-risk for. He had booked a hernia surgery and, before his operation, he learned that individuals with HD needed a specific mixture of anesthetic or they may die during surgery. He presented this information to his doctor, who both knew nothing of the phenomenon and was not interested in learning about it. Keith promptly cancelled his operation and began to look for another surgeon. During the interview, he described his perception on dying in surgery, as

I’ve thought about this since, I mean, what a hell of a way to go. I mean, I’ve see people who had cancer, I mean, I’d much rather die a nice peaceful death than never come out of the surgeons, the last thing I could remember is seeing...counting backwards from a hundred and getting to 97, in retrospect I mean that is not a nice way to go.

Here Keith notes both his risk for an accidental death, as well as his preference for a death from HD, which he suspected would be similar to cancer. As such, Keith emphasizes the non-HD risks to his life, while suggesting that dying from HD may actually be preferable in comparison.
Like Keith, Kate (57, diagnosed in 2005) also contextualized HD within the illness experiences of other people. She noted that there are numerous ailments that did not have a cure, but that did not equate with thinking that your life had preemptively ended. She said,

I think all you can do is try to make the best out of everyday. Live everyday and enjoy it because. At this stage anyhow there’s no remedy, so it’s got to be with your attitude and everybody has a chance. Like my uncle, he was a great believer, he had cancer and lost his foot, and then he lost it at the knee, and that but he always said “You go the, to the hospital, there’s always somebody worse off than you.”

Here Kate suggests that, despite the medical evaluation of the disease, “everybody has a chance” and that individuals with life threatening illnesses need to reflect this in their attitude. Furthermore, as Keith suggested, there are other ways of dying that are comparably worse.

Similarly, another way that death was contextualized by chance was to emphasize other, often more relevant, health concerns. For instance, Celilia (42, gene-positive) noted that HD was not her most pressing concern vis-à-vis her health. She said, “Although I am concerned that I will develop this, there are so many other things that could get you before this. Or could get me, say, before that. To me, the chances of breast cancer to me are a much higher chance then getting this or, living long enough to develop this.” As Cecilia notes, she may have the HD gene, but like many other women of her age group, breast cancer is a much more salient health concern.

Similar to the comments about getting hit by a bus, statements about other health concerns foregrounded other risks in relation to HD. In addition, these were risks that a great number of other people shared, or situations that others were also vulnerable to. Thus, while placing their experience in comparison to the experience of others, these statements also draw on notions of chance to remove the certainty of death from HD from the individual’s potential biography.

The ambiguous nature of being diagnosed with a long-term fatal illness was particularly noticeable amongst those individuals who were asymptomatic, or were only experiencing slight symptoms. These individuals found themselves in the paradoxical position of having a condition
that was fatal, but not yet experiencing any major physiological difficulties. Of all the respondents, Delores (34, gene-positive) perhaps conveyed this experience in the greatest detail as she compared her situation to that of the individuals around her:

> When you tell people it’s not funny, but it’s kind of funny ‘cause it’s almost like you’re telling people that like you’re dying because everyone starts crying and they’re so emotional and they’re so upset which is really sweet because everyone cares so much but it’s like “I’m not dead!” and “I’m not even sick! Not even at all!” [Laughs] You know what I mean? You kind of think “Enough already!” …. The way I kind of see it is... if nothing else gets me, basically Huntington’s will. But you know the truth is – nobody – nobody knows what’s gonna happen. Nobody! His cousin got married this summer, and he just got diagnosed with testicular cancer and my friend who got married five years ago had lymphoma and... my other friend who got married a couple years ago, she had two kids, her husband, 38, had a massive heart attack on his way to work. But I don’t understand – you look at all these people that look like these happy little families, every family has something! Every family has something whether it’s heart or cancer or breast – like look at the breast cancer families. Every family has something.

Delores expressed two common thoughts held by other people with HD: that other families are similarly at-risk of something just as fatal as HD, and that, regardless of the certainties of having or developing HD, no one knows what is going to transpire. While the genetic test for HD may be able to tell an individual that they are going to develop a fatal illness in the course of a normal lifespan, it cannot tell them that they are going to have a normal lifespan. These participants further qualify the results of their diagnosis by emphasizing that their experiences are not unlike the experiences of others. Indeed, they are no longer just “at-risk” for HD, but will develop it, yet what they emphasize, as seen above, is that the only difference is they know what they may die from. The individuals who develop testicular cancer or who die from sudden heart attacks are not somehow immortal in comparison, but rather are unaware of what is likely to end their life. HD seems to place the individual somewhere between “knowing” and “not knowing” in regards to death; as Delores and others suggest, the end of their lives are unknowable, but they do have a possible ending, or as she states, “if nothing else gets me, basically Huntington’s will”.

24
Proximity

While these participants used statements about probability, risk and chance to contextualize death from HD as similar to other health risks and death from other conditions, the aforementioned tension between knowing and not knowing was perceived as unique. That is, while participants were reluctant to view their lives fatalistically, or to even preemptively write its last chapter, knowing one possible outcome, namely death from HD, did change the way that they looked at dying. Specifically, individuals were confronted with their mortality and issues around death much earlier then others. Marvin (60, diagnosed in 2003) said, “But I think we tend to think about things – you know, when it comes to issues like death whereas most people are generally, they don’t even have to deal with those until they’re eighty or ninety.” As Keith (55, diagnosed in 2003) stated, it was as though death was over the horizon line for everyone else, that it was a destination that existed, but was largely unseen, mysterious and not in need of immediate consideration. However, after his diagnosis with HD, death became more tangible. He said,

I realize that I’m going to arrive a lot sooner (at death)...Before, you think you’ll live forever, well I mean, it’s always way far off, that’s the way the average person sees it... Unless you’re really a realist, I guess... So that’s the one thing, you have to confront death maybe a lot sooner than you would have liked.

Thus, while death from HD was just a possible outcome, it was still a very real outcome. In contrast to other individuals, who may face an equal risk, individuals living with HD cannot be oblivious of this potential.

Mark (60, diagnosed in 2009) was perhaps the most poetic in terms of his attitude towards the proximity of death. His perspective had largely been influenced by his father’s suicide, and most dramatically, by his wife’s passing from cancer. Numerous times throughout the interview Mark emphasized that humans were, by nature, finite creatures:
After that experience it is very easy now to see that life is finite. We’re here, but we are finite. It’s like being told you’re pregnant, and you’re to expect a child, but you don’t know when. So until then I’ll do whatever I can to reduce the onset of whatever Huntington’s produces.

Thus, what makes HD unique is the certainty of developing severe and perhaps terminal symptoms, in combination with the uncertainty of knowing when these symptoms will occur. For Mark, the resultant expectancy is much like that a mother experiences, knowing that she will have a child, but unsure of when it will actually arrive.

Phyllis (23, diagnosed in 2008) perhaps best detailed these tensions between chance and certainty and between knowing and not knowing. She had watched her mother live and die with the illness, and she and her siblings had discussed their likelihood of inheriting the gene while waiting to reach the age of majority to take the predictive test. Although she had gone her whole childhood thinking that she would not inherit the mutation, the genetic test determined that she had. In the time leading up to her interview, Phyllis had also received her neurological diagnosis confirming that the disease was beginning to affect her, approximately two decades before HD’s typical symptom onset:

‘Cause whenever I talk to my friend about stuff and I’d like blabber too much and they’re like “yeah” and they’re like “you know what everybody is dying”. I was like, “I know everybody is dying, but everybody is not dying as fast as me”, it’s not the dying part that I don’t like, it’s the fact that it takes so long, that’s the only part that I’m pissed off about. If it took five years, I’d be like “Pph! Whatever, great!” you know? It’s fine, I can do five years. But the fact that it’s fifteen to twenty years and at least 10 of those are going to be spent in a hospital where you’re around and your family will have to come and see you and all your friends and everybody’s going to not know what to say ‘cause they won’t – some days you’ll recognize them, other days you won’t. And so it’s a lot harder to view like dying and everything. It’s one thing just to die, or it’s another thing to even like have it a lot later in life, but when you’re like so youngish too.

Phyllis concedes, as many of the quotes above emphasize, that individuals living with HD are not unique in the sense that everyone is dying. Whether it is from old age, accidents, undetected diseases or HD, everyone is equivalent in the experience of death. The difference that she argues,
a difference that has also been suggested by others, is of proximity. Unlike her peers, at 23-
years-old, she is faced with the possibilities of isolation and loneliness within a care home or
hospital. Phyllis continued her story by discussing her recent accidental pregnancy. Although she
was first concerned about having HD and being pregnant, she eventually decided to keep the
baby. Unfortunately, she experienced a miscarriage. Perhaps the most important part of Phyllis’
account is her juxtaposition of watching her mother pass away with the hope and joy of having a
child of her own. She said,

My mom was 43 when she passed away. She died of starving to death. And it changes a
lot ‘cause her muscles basically shut down ‘cause she said she didn’t want a feeding tube.
I watched my mom die my whole entire life and I’m able to do that. To not know your
own kids, and everything, that is really hard.... But I realized that all I wanted more in
the world was to have that child and even if I could only have five years with it, those
five years would be the most precious and amazing years ever.

In many ways, in contrast to other participants, Phyllis sees a bleak future as certainty and
believes that she will die in a manner similar to her mother. However, it is important to note that
she still refuses to truncate or finalize what she suspects will be the last relatively unaffected five
years of her life. Despite being aware of the realities of dying with HD, and growing up with a
parent in palliative care, Phyllis emphasizes that both she and her unborn child would still have a
life worth living.

While Phyllis’ account does contain elements of bleak certainty, it also highlights her
remaining opportunities. She, like many of this study’s participants, acknowledged that a
diagnosis of HD creates a more imminent relationship with death. However, these participants
also emphasized that their future is not certain. As these participants noted, having a fatal illness
does not equate dying from that illness, nor does their death begin at the time of diagnosis.
Suicide

Suicide was not a particularly common topic within the interviews\(^3\). As shown, individuals typically avoided fatalistic statements about death and were also quite optimistic about a cure for HD within their lifetime. That said, several individuals had experienced episodes of suicidal thoughts, or were suspected by others to be contemplating suicide.

Two participants in this study openly discussed their own actions in relation to suicide. The first, Phyllis (23, diagnosed in 2008), recalled the most recent occasion that she had attempted to take her own life. Prior to her confirmatory genetic test, Phyllis was certain that she would not inherit the gene for HD. To her disappointment, her test came back gene-positive. In what Phyllis thought was an ironic twist, her sister, who had feared that she would inherit the gene since she was young, had received a gene-negative result. Their relationship with one another switched, with Phyllis' sister now acting as the caregiver. However, despite her attempts to maintain optimism, Phyllis became depressed and suicidal. She moved away from her family to Vancouver, where she struggled to meet new friends and start romantic relationships under the shadow of HD and the uncertainties around disclosure. Phyllis decided to attend a party in a neighboring resort city with some new acquaintances, but decided to leave the party on her own as a result of her feelings of alienation. In a decision that echoes the actions of Phebe Hedges (Wexler, 2008), Phyllis decided to remove her winter clothing and lay down in the snow:

So I went walking to this lake 'cause I had a plan to fall – just lie down in the snow so I wouldn't have to get up anymore and deal with anything. Then I was lying there in the snow thinking “what am I doing?” after a bit, “I shouldn't be doing this; I am such a frickin moron!” And it was really cold out, and I managed to lose my mitts and any toque and everything. So I realized this after a bit of time of lying out there that I was like, “ah, this is stupid” so I got up and I was like stumbling through the snow trying to get back to like the road kind of thing. And then, I fell a couple of times along the way and then the last time I just couldn't get up anymore 'cause my body was like so like cold and frozen. And I was lying there and I could seriously – it was just disgusting – my hands and everything were just like frozen, it was gross. And I was staring up praying like

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\(^3\) Please refer to the Appendix 1 for a discussion of the issues of interviewing participants about suicide.
desperately “Oh God, what have I got myself into? I can’t do this, I’m so stupid!” And I’m like saying goodbye and then all of a sudden my hands were instantaneously warm. And I was like – my God! Freaked out, jumped up, ran through the snow and there was a car so I like hailed it, and it was a police car and they took me back to the housing place where I was staying and then they got mad at me for telling them a story [laughs]. Yeah! And then, “You shouldn’t be telling stories like this!” blah blah blah and I was like “I’m sorry, I was drinking too much, got lost and wanted a lift back up” they were like “That’s better! Now don’t go doing this again!”

Although Phyllis mentioned that she had attempted suicide previously, and in this case, she decides that she has made a mistake after she places herself in a potentially dangerous and vulnerable situation. Fortunately, Phyllis was able to remove herself from her circumstances and secure a safe ride from the police before she seriously injured herself. The reaction of the police to her story is quite noteworthy. Rather than accept her attempts to inform them that she has attempted suicide, they coerce her into reworking her narrative into a more palatable story, that of a party misadventure.

In addition to Phyllis, suicide was also particularly salient for another participant. He contextualized his perspective by recounting his experiences of growing up in a very religious environment. As he grew older, he began to reject many of his childhood teachings and became highly identified as an atheist. Before the recorders were turned on for our interview, he asked me vague questions about my beliefs, in an attempt to ascertain any religious affiliation. He also asked that the recorders be turned off for conversations about a “particular topic”. This topic was eventually revealed to be his attitude on suicide and he was particularly concerned with how the recording could affect aspects of his future life. Accordingly, these sections of the conversation have been withheld4. He did, however, give permission to present his more general perspective on suicide. He said:

I’m very easygoing about the end of life. And my wife is also, because she is [ethnicity] and it’s very common over there to arrange your death, you know at the end. And her uncle did just that. And I knew him well. He was a doctor. Actually her uncle and myself

4 The participant’s pseudonym and basic demographics are also detached from this quote.
were very similar. We were very secular. We kind of... you know got along well. And one time we were out for a walk and I told him I would, if I got really sick, you know at the end of my life, I would – by then I had known about HD for five years, so I was talking to him about stuff like that. And he understood. He could read about it and find out. So I told him that if it became a problem for me I would also – I would do that. Ah, it’s not a big deal. Because my attitude is like life is like a gift everyday. Every day is like a free day to me. And not every day is a good day and not every day is equal but – in general it’s pretty good. So... I am not worried about that kind of thing later on. I think as long as I’m physically okay, that’s good. But if I start to go mentally, I will probably think that’s very depressing. And I’ll probably be depressed by that whole idea because my whole being is very mental oriented in a way. That’s sort of how I see life, and I’ve had all these nice mental thinking days of my life. And I regard my religious days as you know –not really fully living – I was living in fear of funny things. Superstitious things.

This respondent’s perspective on suicide was intimately connected with his personal identity. For instance, he avidly rejected the religious teachings of his youth and gravitated to “rationality”, which would not stigmatize an action such as suicide. He was also particularly concerned about, and invested in, his mental capacities. The deterioration of his mind was accordingly connected with a decreasing desire to stay alive. It is also interesting to note, despite the participant’s engagement with suicide, that he does not use a term such as “ending my life” or “suicide” in connection with himself. Instead, he recalls what his relative did and states, “I would do that”. As such, despite his matter-of-fact discussion of suicide, there still seems to be a distance between himself and the act. He then told the story of a relative suicide, noting the benefits of suicide:

So he had this good evening with his brother, kind of a goodbye evening. He went home and said “I’ve had a great evening this evening”, he wrote out a little note. He said, “I’ve decided now to, to end my life and on a nice evening such as this and I have the proper medicines and I’ll have no pain” and he did it himself. He said, “I would just like you to come and find me” um “or call the authorities” you know, “I’ll be at home here” and what he did was he mailed the letter. So that’s what happened. His brother didn’t open the letter until the following day of course, then – you know, but he realized of course his brother was just laying in the bed dead, no problem there. So that’s how it went. And then you know he asked [his brother] to explain it all to the family and it’s no problem at all. Nobody was the least bit –everybody was kind of happy about it actually.

Through the story of his relative, he emphasizes the positive and desirable aspects of suicide. As this participant presents it, his relative’s suicide made all those around him happy, while ending
his own suffering. It is of particular interest that the participant does not include any accounts of grief or loss that the family members felt at their loved one’s passing.

Other individuals’ experiences of suicide were mediated through their contact with health professionals, who suspected that they might consider taking their own life. In particular, the period directly after diagnosis was extremely depressing for several individuals. One participant, Gerald (52, diagnosed in 1996), stated that his diagnosis with HD felt like a “death sentence”, and it took him several months to regain his vigor for life. This improvement was partially connected to his regular appointments with a therapist, whom he had started seeing after he was diagnosed.

Similarly, other health professionals attempted to arrange more frequent sessions with individuals they suspected may be vulnerable to suicidal thoughts. For instance, Mark (60, diagnosed in 2009) was booked for sessions with a health professional every six weeks after his diagnosis. He explained that this was contextualized by his father’s suicide, even though he stated that he would never consider taking a similar course of action. He said, “I told [the doctor], that as a survivor of someone who committed suicide, I would never do it.” Samuel (47, diagnosed in 2003) also had a history of suicide within his family, and health professionals accordingly expressed similar concerns about his possible actions. He said,

I guess they were worried about me killing myself out of there – when they find out the test results and things. They had a form to go through. I don’t know, they use it to determine your suicide skill or something like that. I did for a while, after a while I was -- actually, I pretty well gave up on things.

Both Mark and Samuel noted that, in addition to their family history of suicide, they were quite depressed at the time of their diagnosis. Unlike Mark, Samuel acknowledges that he did initially consider suicide, and although neither of them stated that they abstained from suicide because of these actions, they both appreciated the concern that health professionals had for their lives.
It is also noteworthy that neither Mark nor Samuel stigmatized their father's actions. While they decided that suicide was not an option for them, and Mark even noted that he was a "survivor" of suicide, they could still empathize with the choices made by their fathers. Samuel discussed this understanding, as he detailed his father’s suicide, which occurred when he was 12-years-old.

We had a farm and, and one day he [father] just kind of came out, came over and said, “Okay, we’re going to go and put” – well – he didn’t tell us, actually he just brought in the veterinarian and he put all the animals to sleep on the farm. I guess he felt maybe we couldn’t take care of them. I guess. Another day, like he took me aside and said, he just made sure I knew how to run the farm, and stuff like that, you know. And made sure I had all these things on track. And – of course, what happened – he killed himself. But when you’re sick, when you are an adult you suddenly realize how difficult decisions are, when you have Huntington’s and stuff like that too. So things that didn’t make a lot of sense back then tend to be a little clearer as an adult. I always felt it was a good decision. He had a mum in the hospital for 30 years and I guess it was a big commitment on the family; I’m sure for him. To go visit her all the time and she was quite shaky and things. It wasn’t a very bright future for Huntington’s at the time of the passing. So I can certainly understand why he made that decision and I think he really kind of did it for our family’s benefit too.

Samuel’s father’s actions distressed him greatly when he was younger. However, as he notes in this account, he now has a new clarity on his father’s actions. Like the unnamed participant above, he feels that his father’s suicide was for the benefit of the family. He also states that he personally thinks that his father made a good decision, as well as noting how difficult it is to make decisions in the position that his father was in. Thus, despite having a family member who took their own life, both Mark and Samuel express sympathy and understanding for individuals who commit suicide.

Other individuals within the HD community expressed a similar attitude towards suicide, one that did not perceive suicide as stigmatizing and expressed empathy for the individual who chose to end their own life. Donna (47, diagnosed in 1999) conveyed such sentiments as she recounted her frank conversations with several individuals who were considering suicide. She said,
There are people who are not happy in life, also at times. Some of them, it’s a major issue. I know a couple of people that contemplate committing suicide. Towards the end of my first marriage I was really depressed, just like crying everyday. Then I just understood how desperate people would feel, who commit suicide. But that’s not something I’ve ever thought of doing myself. I’m enjoying my life too much [laughs]. I always try to stay positive for people. You can’t change people’s minds though.

Like both Mark and Samuel, Donna expresses understanding for individuals who are considering taking their own lives. In particular, she relates their situation to her own period of extreme depression. While she does not consider suicide an option for herself, she does emphasize that it is an individual’s choice.

The issue of stigma becomes particularly noteworthy when comparing the accounts of the two individuals who are considering suicide to those individuals who state that they would not take their lives. As mentioned, the unnamed participant avoids connecting any verb for suicide to himself, and instead speaks about the act in vague terms. He also expressed a great deal of hesitation about the topic, and wanted to ensure that the researcher was not judgmental before the topic was raised. Similarly, Phyllis calls herself a “frickin moron” for her attempt, and when she reveals it to police officers she is pressured into creating a more palatable story. However, other members in the HD community, including those with specific suicide experience, do not stigmatize suicide. Indeed, they sympathize and understand why a fellow community member would take such a course of action. Individuals such as Donna also highlight that suicide is a personal decision, stating that you “can’t change someone’s mind”. She, along with the health professionals mentioned above, attempt to support individuals who appear suicidal. However, just as importantly, Donna and the other participants refrain from making normative judgments on those individuals with HD who are considering, or have committed, suicide.
Conclusion

These sections present contrasting perspectives. A few individuals with HD actively contemplated suicide, while the majority are resistant to fatalistic interpretations of their lives. However, as far as HD research is concerned, suicide is the most salient issue vis-à-vis death (Farrar, 1986; Paulsen, 2001; Paulsen et al., 2005). Obviously, the risk of suicide in relation to predictive testing is of great concern for those health professionals who administer such tests. However, it is also important to qualify the reality of such risks with the perspectives displayed by the majority respondents in this chapter. That is, while they may have a fatal condition, their lives have not been finalized by that condition and suicide is subsequently not considered a course of action.

As has been noted, the experiences of individuals suffering from degenerative diseases are not well accounted for within the literature (Exley, 2004). Instead, the majority of research has focused on individuals in the terminal stages of disease and those already in hospital care (Glaser & Strauss, 1965; Illich, 1975; Lawton, 1998; Seale 1998; 2000; Sudnow, 1967). The two studies that discuss HD and palliative experiences (Dawson et al., 2004; Roger, 2007) similarly focus on this stage of advanced illness. However, the perspectives of individuals living with HD in this chapter suggest that death can be a salient issue well before the physiological experience of dying begins. Furthermore, their views on death were quite different than those reported by both Dawson et al. (2004) and Roger (2007). While both of these reports emphasized a fear of the future, the participants in this study did not express comparable apprehension. In contrast, their accounts of suicide as a personal decision, as well as their refusal to allow their diagnosis of a fatal condition to finalize their lives, emphasized their desire to maintain their agency and avoid the preemptive abbreviation of their biographies. As such, by extending the period of analysis outside of the terminal stages, important nuances in the individual’s relation to their death may be obtained. Furthermore, although they were living with a fatal condition,
respondents also noted the mortality of all individuals. They often stated that there were
similarities to other diseases, other forms of death, and human life in general.

These statements echo Simmel’s (2007) perspective on death, particularly his notion that
mortality is a defining human quality. Whether we acknowledge, or deny it, our existence as
living beings is partially defined by the fact that we all die. As Simmel states, “death and life are
intimately conjoined…. Death limits, that is, it gives form to life, not just in the hour of death,
but also in continually colouring all of life’s contents” (p.74). Subsequently, what both Simmel
and these participants underscore is that we are all mortal. Respondents’ experiences with the
symptoms, degeneration and disability of HD may be unique to that disease but, as they remind
us, we all share a common end. However, as numerous authors have noted, social research has
avoided or negated issues of both the body and mortality (Frank, 1995; Hawthorne & Leaman,
2001; Schilling, 1995). As medical technology advances, an increasing number of individuals
will become cognizant of risks to their lives well before physiological effects appear, similar to
those experienced by these respondents (Davidson, MacIntyre & Smith, 2008). Accordingly, as
social research must become more aware of the body and mortality, investigations of death and
dying arguably need to consider how the phenomenon is experienced before it enters the stages
of conspicuous embodiment.

Having discussed issues relating to death and suicide, this thesis will now turn to
individual’s experiences with HD symptoms. Specifically, the subsequent chapter will focus on
the psychiatric aspect of HD and the misdiagnosis of HD as a mental disorder.
CHAPTER 2: THE PSYCHIATRIC MISDIAGNOSIS OF HUNTINGTON DISEASE

Introduction

A precise genetic test has existed for HD since 1993. However, this test only identifies the individual’s number of CAG repeats, and therefore, only the potential to develop the illness. Diagnosis of actual disease onset can only be made by a neurologist, based on the detection of tangible changes in the individual’s brain and motor symptoms (Rosenblatt et al., 1999). However, even though HD has a number of symptoms associated with neurological ailments (e.g., chorea), it also has several others that may appear to be psychiatric in origin. For instance, individuals with HD might notice changes in their mood and personality, as well as experience hallucinations (Paulsen, 2004; Yu, 2004). Furthermore, recent reports have suggested that the psychiatric and cognitive changes associated with HD can appear years before the characteristic neurological changes (Diamond et al., 1992; Duff, Paulsen, Beglinger, Langbehn & Stout, 2007; Foroud et al., 1995; Hahn-Barma et al., 1998; Jason et al., 1988; Johnson et al., 2007; Kirkwood et al., 1999; Langbehn & Paulsen, 2007; Marshall et al., 2007; Morris, 1991; Snowden et al., 2002; Solomon et al., 2007).

As such, even though there is an accurate genetic screening test for HD, the disease’s symptomology and progression can make accurate diagnosis a complex process, particularly so given the estimated 25% of individuals with HD who additionally lack any family history of the disease (Almqvist, Elterman, MacLoed & Hayden, 2001). The following chapter will suggest that the interpretation of HD’s features within psychiatry’s epistemology highlights ambiguities between psychiatry and neurology. However, this ambiguity is more than a simple epistemological difference, as it also has tangible effects. These effects are evident in numerous case study reports on the psychiatric misdiagnosis of HD (e.g., Jardri et al., 2007; Yu, 2004).

Several less accurate tests preceded the one referred to here.
Although these case studies do detail the misdiagnosis of one or two individuals with HD from a clinical perspective, no published study has yet engaged with the HD community regarding psychiatric misdiagnosis. This chapter will address this absence by highlighting the accounts of individuals currently living with HD. Their recollections of familial misdiagnosis, and, in one case, a personal misdiagnosis, will be used to elucidate the individual consequences of the disjuncture between neurology and psychiatry. After tracing both this epistemological tension and its consequences, this chapter will discuss these findings via Latour’s (1987; 2005) concepts of oligopticons and metrology, as well as Bakhtin’s (1986) notion of speech genres.

**Diagnosis, Misdiagnosis and Psychiatric Epistemology**

Although there have been no previous empirical articles on the psychiatric misdiagnosis of HD, the accuracy of psychiatry has been questioned from numerous perspectives. Most famous amongst these critiques is Rosenhan’s (1973) article, which questioned psychiatry’s ability to distinguish the sane from the insane by documenting the diagnosis and psychiatric hospitalization of several healthy individuals. Other researchers have raised questions over psychiatry’s cross-cultural applicability (Good, 1993), the accuracy of its diagnostic categories (Greenberg, 1977), the repercussions of its internal divisions on the selection of diagnoses (Brown, 1987; 1990), the disjuncture between its medical training and in situ experiences (Thomas-MacLean & Stoppard, 2004), and the fabrication and alteration of diagnoses by psychiatrists (Pallone & Hennesy, 1994).

There are also numerous commentaries on the shaping of psychiatric epistemology by larger social narratives. For instance, andocentric assumptions about the nature of women’s psychology have been connected to the misdiagnoses and inaccurate treatment of women by psychiatric clinicians (Floyd, 1997). Most prominently, Smith (1975; 1978; 1983) has explicated the problems psychiatry has in both understanding women and interpreting their experiences.
Other researchers who have noted psychiatry’s inattention to social factors affecting diagnoses include Sayre (2000), who discussed the lack of regard clinicians afford to the patient’s living situations pre-admission and Jarvis’ (2007), who highlighted a similar inattention to the connections between psychosis and social factors, such as poverty, migration and race. These limitations of psychiatric epistemology and diagnostic processes have been connected to numerous social outcomes, such as the well-documented overrepresentation of mental illness diagnoses in minorities in the United States and Great Britain, particularly in reference to schizophrenia and migrant populations (e.g., McKenzie, 1999; Neighbors, Trierweiler, Ford & Muroff, 2003).

Recent research does, however, indicate that psychiatrists have begun to react to these external criticisms and are cognizant of the ambiguities of their epistemology. For instance, Messinger’s (2007) ethnographic research described the negotiated construction of psychiatric diagnoses by an interdisciplinary team, as well as the solicitation of input from non-psychiatrists in an urban hospital. Additionally, Rafalovich’s (2005) report documented the ability of clinicians to negotiate the controversial nature of ADHD diagnosis and treatment. He also highlighted how clinicians maintained a skeptical attitude toward ADHD, whilst exercising their autonomy in the treatment process. These reports highlight some of the novel avenues used by clinicians to ameliorate some of the omissions and contradictions in psychiatric epistemology.

Within the context of this chapter, the questioning of the epistemology and diagnostic ability of psychiatry is backgrounded by the numerous difficulties already occurring after an individual has been diagnosed with a degenerative illness. For example, a diagnosis can initiate a dilemma between the degenerative nature of the illness and a desire to maintain self-identity (Harman & Clare, 2006), or the family of the diagnosed individual might experience caregiver burden paired with grief (Morgan & Laing, 1993). A number of these difficulties are specific to the context of HD. For instance, individuals can face stigma based on their movements (Paulsen,
2004), as well as employment and insurance discrimination over their genetic status (Bombard et al., 2007; 2008). The familial and hereditary nature of HD also places the family in a unique situation, as they struggle with communicating genetic risk (e.g., Cox & McKellin, 1999). Thus, an accurate diagnosis of a neurological condition, such as HD, carries with it major implications for both the individual and their loved ones. A misdiagnosis delays the onset of encountering these issues, as well as lessening the time individuals can be engaged with them. Psychiatric misdiagnosis also carries consequences of its own, as individuals with psychiatric illness also often face stigmatization (Sayre, 2000) and might develop psychological problems in response to their misdiagnosis (Floyd, 1997).

In summary, there have been numerous critiques of the epistemology of psychiatry. Many of these critiques can be connected to tangible consequences, most commonly in the form of diagnostic trends and misdiagnoses. However, recent reports suggest responsiveness in psychiatric practice to some of these issues. The following analysis will synthesize these elements in the following manner.

**Neurology, Psychiatry, and Huntington Disease**

There have been numerous reports indicating that the psychiatric components of HD can, in fact, be the first symptoms to appear. For instance, Duff et al.’s (2007) study of 681 individuals with HD from America, Australia and Canada suggests that psychiatric symptoms appear well before the cognitive and neurological markers of the illness, with emotional dysfunction often being the first symptom. A similar study observed that, compared to controls, a Huntington gene positive cohort had greater difficulty recognizing other’s negative emotions; this difficulty increased as they drew closer to their eventual neurological diagnosis (Johnson et al., 2007). Additionally, a report by Stout et al. (2007) suggests that cognitive decline also escalates well before the neurological diagnosis, and, similar to the emotional effects of HD, can be an “advance warning”
of motor symptom onset. Thus, initial psychiatric effects, followed by a more pronounced chorea, constitute the trajectory suggested in the findings of several recent studies on HD.

This literature on the early onset psychiatric effects of HD indicates how the disease can blur the boundaries between “neurological illness” and “psychiatric illness”. While the disease itself can manifest as a hybrid of neurological and psychiatric symptoms, the institutions and disciplines of neurology and psychiatry are not as seamlessly interwoven. Although neurologists and psychiatrists both receive training as medical doctors, and share a similar organ of interest, the brain, the perspectives that they bring to the study of this organ are quite different.

Accordingly, psychiatrists and neurologists have noted the tension between their two perspectives (e.g., Cunningham, et al. 2006; Schon, MacKay, & Fernandez 2006a; 2006b; Selwa, Hales, & Kanner 2006). A trio of neurologists, Price, Adams and Coyle (2000), most directly summarized this tension as a “great divide”. This divide centers around the neurological tendency to focus on the “brain” compared to the psychiatric tendency to focus on the “mind”. Although these disciplines have been similar but separate throughout their histories, it is precisely diseases such as HD that are creating the need for greater interchange between them, leading to the authors’ suggestions that these two professions need to receive a more harmonious education (Price et al., 2000). Even though Price et al. (2000) and authors of similar articles (e.g., Cunningham, et al. 2006; Schon et al., 2006a; 2006b) underscore both the overlaps and disjunctures between neurology and psychiatry, their focus remains primarily on epistemology and pedagogy. However, these different conceptualizations of the mind/brain have repercussions outside of academia, as their operationalization creates tangible consequences for clinician and patient, consequences that are especially evident in the misdiagnosis of HD as a psychiatric illness.

A review of the case study literature on such misdiagnoses uncovers multiple instances in which psychiatrists have mistaken HD for a mental disorder. For instance, Jardri et al. (2007)
present the case of “Oliver”, who was diagnosed with Major Depressive Disorder with Psychotic Features, but did not respond to appropriate treatments. It was not until HD was discovered in Oliver’s family that his treatment was adjusted and his condition improved. Similarly, Duesterhus et al. (2004) also present a case study of a severely depressed child, whose diagnosis was informed by a long-standing family history of Major Depressive Disorder. Of particular interest is the child’s father’s diagnosis with Major Depressive Disorder with Psychotic Features; it was not until his eventual suicide attempt that his HD was detected, subsequently explaining the family’s apparent depressive genealogy. As with Oliver, this child and his father’s condition greatly improved once they and their healthcare providers were made aware of their actual condition. In both these case studies, psychiatrists noted that the hereditary depression was a “red herring”, concluding that psychiatrists and mental health professionals needed to be more sensitive to HD.

The psychiatric record of the misdiagnosis of HD extends far past these case studies. Appollonio et al. (1997), in their report of three cases of misdiagnosed HD, suggested that genetic tests and genealogical histories be included within standard operating procedures for individuals with symptoms similar to HD. Yu (2004) echoed these statements in her report on the lack of screening for HD, also noting its prima face similarity to first break schizophrenia. She also estimated that psychiatric disturbances account for as much as 25-80% of the first symptoms of HD. Tost and colleagues (2004) concurred with these suggestions in their report of a man who had been diagnosed six times with four different conditions over the course of seven years before substantial chorea appeared and he was tested for HD. Even though this man’s condition stabilized after the correct diagnosis, he had already taken anti-psychotic medications for multiple years, and had been both homeless and incarcerated numerous times as a result of the misperception of his psychiatric symptoms.
These psychiatric case histories provide ample evidence of the difficulties in diagnosing HD, as well as the tangible consequences of the disjuncture between psychiatry and neurology. It is also noteworthy that all of these misdiagnoses were made after the development of the genetic test for HD. Although this test is an accurate tool for detecting HD, the clinician first needs to suspect HD as a possible diagnosis. However, it is perhaps most disappointing that each of the aforementioned articles echoes statements made in Stewart’s 1987 report on the psychiatric misdiagnosis of HD, which appeared in the *Journal of Neuropsychiatry and Clinical Neuroscience*, the official journal of the American Neuropsychiatric Association. At least one decade prior to all of the previously discussed case studies, in a journal focused on neurology and psychiatry, Stewart (1987) outlined the risk of misdiagnosis and forwarded a protocol stating that individuals who meet two of the three HD symptom criteria should have the disease eliminated as a possibility through differential diagnosis. Despite this timely and well placed advice, the lessons from Stewart’s report on misdiagnosis apparently did not cross the “great divide” and alter the outcomes for the individuals or the practices of the clinicians in the aforementioned case studies.

At this point, two important factors become clear. First, HD has numerous psychiatric symptoms, which might appear prior to the characteristic chorea. Second, given the psychiatric nature of HD, it is quite possible that an affected individual’s first point of contact with the healthcare system will be made through a mental health professional, as demonstrated by the numerous case studies reviewed above. As such, there is a risk of the individual being misdiagnosed with a mental disorder, rather than HD. Given the overlap of symptoms across HD and other mental illnesses, it is not surprising that mental health professionals make these errors; however, it is surprising that there is such a proliferation of case studies repeating the same accounts of the risks of misdiagnosis and suggestions to avoid similar mistakes. Subsequently, it
is arguable that the seemingly ambiguous nature of HD needs to be communicated to psychiatrists via a better means than journal case studies.

It is here that one must turn to the orientating text of psychiatry and mental health in North America, *The Diagnostic and Statistical Manual of Mental Disorders* (DSM), which is currently in the revised version of its fourth edition (APA, 2000). The DSM-IV is the text that organizes, describes and standardizes mental illnesses. Given that the DSM-IV is a pivotal text for providing accurate and uniform descriptions of mental pathologies, paired with the string of literature on HD misdiagnosis, it would seem the logical place for a concise and clear definition of the psychiatric components of HD. Below is the complete DSM-IV content relating to HD:

The essential feature of dementia due to Huntington’s Disease is the presence of a dementia that is judged to be the direct pathophysiological consequence of Huntington’s Disease. Huntington’s Disease is an inherited progressive degenerative disease of cognition, emotion and movement. The disease affects men and women equally and is transmitted by a single autosomal dominant gene on the short arm of chromosome four. The disease is usually diagnosed in the late thirties to early forties but may begin as early as age four years in the juvenile form or as late as age 85 years in the late onset form. The onset of Huntington’s Disease is often heralded by insidious changes in behaviour and personality, including depression, irritability and anxiety. Some individuals present with abnormalities of movement that resemble increased fidgeting and that later progress to characteristic generalized choreoathetosis. Difficulties with memory retrieval, executive functioning, judgment are common early in the course, with more severe memory deficits occurring as the disease progresses. Disorganized speech and psychotic features are sometimes present. Late in the disease, characteristic “boxcar ventricles” may be seen on structural brain imaging due to the atrophy of the striatum. Positron emission tomography (PET) may show striatal hypometabolism early in the disease. Offspring of individuals with Huntington’s Disease have a 50% chance of developing the disease. A genetic test is available to determine with relative certainty whether a given at-risk individual is likely to develop the disease; however, such testing may be best administered by centers with experience in counseling and follow-up of individuals at-risk with individuals at-risk for Huntington’s Disease (APA, 2000, p.165).

This codification provides a number of important and useful criteria; for instance, it highlights all three primary areas of HD (psychiatric, cognitive and motor) whilst portraying a descriptive account of individuals who might be presenting with HD. Yet the account, titled “Dementia due to Huntington Disease”, is primarily included to attune psychiatrists dealing with an individual
who has already been diagnosed with HD that dementia might occur. Even though the entry states that emotional and cognitive changes might “herald” the start of HD, it does not state that HD can be mistaken for other mental disorders with these specific symptoms. Of course, even if the entry did include these details, it would remain of limited use, as the disease is listed as a subtype of dementia, rather than as its own category or being discussed within the differential diagnosis for other pathologies.

The HD entry within the DSM-IV does not convey any of the lessons learned by the numerous psychiatrists who have misdiagnosed the disease over the years. Furthermore, HD is not listed as a differential diagnosis under any other illness category, including Major Depressive Disorder or Schizophrenia, despite the numerous case studies where the disease has been mistaken as one of these other pathologies. Perhaps most telling is that the description of HD labels it as a “progressive degenerative disease” that affects mood, rather than a psychiatric or neuropsychiatric disease. Given the brief presentation of HD with the DSM-IV, as well as the omission of any psychiatrist’s misdiagnosis experiences, it is perhaps understandable that psychiatrists have repeated both each other’s mistakes and each other’s advice. Indeed, the brief inclusion of HD within the DSM-IV is perhaps directly indicative of the disjuncture between the epistemologies of neurology and psychiatry.

The brief DSM entry, the aforementioned case studies, discussions within the neurology and psychiatry communities, and the symptomology of HD itself all illuminate an epistemological gap between neurology and psychiatry. However, omitted from these accounts are the consequences that misdiagnoses have for individuals with HD and their families. The following analysis will place the experiences and recollections of misdiagnosis from individuals with HD in dialogue with this case study literature to further elucidate the tangible consequences of the epistemological disjuncture between neurology and psychiatry.
Experiences and Recollections of Misdiagnosis

When neurologists and psychiatrists discuss the overlaps and disjunctures of their respective positions, their focus often remains on the realms of pedagogy and epistemology (e.g., Price et al., 2000). These debates become somewhat grounded within the psychiatric case history literature, which illuminates the effects such epistemological differences can have. At the same time, omitted from both of these dialogues, as well as the literature on early expression of the psychiatric symptoms of HD (e.g., Duff et al., 2007; Johnson et al., 2007; Solomon et al., 2007), are the consequences that such disciplinary tensions and misdiagnoses have on the individuals to whom they are actually applied. Although no report has thus far highlighted these experiences, the impacts and lingering effects of misdiagnosis becomes evident through the accounts of the HD community.

While relaying stories of their family history of HD, participants in this study frequently recalled cases of familial psychiatric misdiagnosis, as well as highlighting a general culture of HD misdiagnosis in generations past. Participants most commonly expressed both the historical misdiagnosis of HD and the attitude toward the resulting treatment with the phrase “locked up in Thorn Hill⁶”. In this statement, “Thorn Hill” refers to the psychiatric hospital where many individuals with HD were relegated when their illness was incorrectly perceived as being of psychiatric origin. Keith (55, diagnosed in 2003) noted this when he spoke of his father’s treatment: “My father died in Thorn Hill in ’63, but we thought he had schizophrenia. If you look at the family tree there’s a lot of people dying earlier than they should have.” Several other participants who had conducted archival family histories recalled similar misdiagnoses. For instance, Kenneth (76, diagnosed in 2003) stated that, “I’ve done a lot of genealogy and you have to get the records from the psychiatric hospitals because that’s where they were all put. Because they were all thought to be crazy.”

⁶ The name of this hospital has been altered.
Often the consequences of previous familial misdiagnoses were most directly felt when individuals learned that generations of mysterious illnesses or bizarre behaviors were, in fact, HD. For instance, one participant compared the discovery of HD in her family to a “flood” that consumed her family tree:

I had no family history, which as you know is quite odd given it's a genetic thing (laughs). Basically my dad got diagnosed in 2007. He was having some problems with motor skills, but not really chorea, but more so mental issues. He was having a problem keeping a job, etcetera, etcetera, and the last test that they did was for Huntington’s. Just to rule it out. And it came back positive. And he’s got seven siblings who all have kids and me and my sister have kids, so it was like this flood up through our family tree and now like half of it is gone (Delores, 34, gene positive).

The clearest implication of this account is the shock created by the sudden knowledge of a genetic disease within the family. Additionally, prior knowledge of a familial history of HD might have potentially altered reproductive decisions of several of the aforementioned family members. However, as both family members and health professionals alike suspected the family was affected by a disease that was psychiatric in origin, this possibility was not considered.

Other families had similar experiences based on their own histories of erroneous diagnosis. Particularly problematic was when a parent’s misdiagnosis also formed the basis of a child’s subsequent misdiagnosis. Kate (57, diagnosed in 2005) said,

He [her father] had been under psychiatric care, when my mom and him got separated that’s when he had a “nervous breakdown” and that’s when he had been going to a psychiatrist. But it was the Huntington’s and nobody knew. That’s when they put him out to the crazy clinic and he was probably out there for four or five months and they ended up giving him shock treatments to bring him back because he would just sit there. But they had no idea that it was Huntington’s... and my brother got sick right after my father died and they thought it was depression. Which was what my father was misdiagnosed with as well. Well, we were told he had a nervous breakdown.

Here, in an unfortunate irony, it is the father’s apparent psychiatric condition that is seen as the hereditary fact explaining his son’s similar behaviors. Following that misdiagnosis, Kate’s
brother received inaccurate and inadequate treatment, and spent several years of his life in extreme poverty. Indeed, it was not until Kate herself was tested for HD that her father and brother’s experiences were put into an accurate context, and the latter began to receive proper health coverage and disability benefits.

Keith recounted a similar situation, which included the profound consequences of misdiagnosis, stating that his brother, like his father, had his behavioral changes misinterpreted as mental disorder. Keith’s brother was believed to have a psychiatric illness, and, as a result, did not receive appropriate or adequate care, spending most of the last years of his life living on the streets of a metropolitan Canadian city. As Keith succinctly put it, “my brother would probably still be alive if he hadn’t gone undiagnosed [with HD].”

Another participant, Becky (36, diagnosed in 2007), was not misdiagnosed per se, but was receiving psychiatric counseling for an eating disorder immediately before her diagnosis of HD. At this time, she had already developed gait and speech changes. Although her clinician did not suspect a neurological disease, her family members convinced her to undergo neurological testing in a nearby city. Becky went into the assessment to consider the possibility of early onset Alzheimer’s Disease (AD), which her father had been diagnosed with. Upon her visit with the neurologist, HD was almost immediately suggested based on her gait. This diagnosis turned out to be correct and her father’s, as well as several previous generations of AD diagnoses, were invalidated.

Shortly after this diagnosis, Becky developed hallucinations and delusions. At first she suspected she was being watched from across the street. Later, she believed there were cameras in her ceiling. A neighbor later reported overhearing Becky shout to invisible intruders that she was going to “skin them alive” if they entered her home. During our interview, she also recounted her first experience of hearing these voices, as well as her current treatment. She said,
I was with my aunt and she took me out for Asian food. And I thought someone said I was drunk. But there was no one there. I didn’t really think about it, because I didn’t really know, so it took some time... I hear voices and living with it is tough, [inaudible] I’m a psychopath... But it’s a hard thing to explain, it’s a really hard thing. I could hurt people, or hurt myself. But now I’m on the right pills. They told me it’s really important to treat it right away, because it can affect my brain. So it’s very important to have the right pill. But they make me feel very tired, all day long.

Becky’s experiences outline two important points. First, even though clinician did not misdiagnose her, she was receiving mental health treatment after the onset of HD. During this period, her movements and speech were affected significantly enough for her neurologist to suspect HD prior to conducting any formal examinations. However, her clinician neither addressed these symptoms, nor recommended their evaluation by another health professional. Second, Becky’s account also highlights the importance of accurate and timely diagnosis. Indeed, it is not difficult to imagine what her possible misdiagnoses might have been had she developed these hallucinations before receiving the HD diagnosis.

Each of these participants’ experiences documents an instance where the nuances of HD have formed the basis for a misdiagnosis. The repercussions of HD’s ambiguities were perhaps felt most directly by Grace (53, diagnosed in 2005), who was personally misdiagnosed. Grace suspected that she was sick well before her diagnosis with HD, and her ex-husband had also long suspected that she had a serious medical condition, as she recalled him stating, “you’ve got movements and you’re always running around. You’re hyper and you’re going to have a serious disease”. Critically, both of their opinions were primarily informed by Grace’s history of awkward movements: “I’ve always dropped stuff. I’ve always had trouble finding things and the computer part of my brain just wasn’t there and that type of thing.” In this instance, unlike many of the previous accounts, it was actually a neurological symptom that presented as the first HD feature detected by both the individual and their family. However, it was not her movements, but rather issues emerging related to her marital difficulties that influenced Grace to seek the help of
a psychiatrist. Her psychiatrist prescribed her several medications, including a powerful anti-psychotic medication (Haldol) typically used for people with acute psychosis and schizophrenia. Eventually, given her repetitive and awkward movements, both Alice and her psychiatrist surmised that she had Tourette Syndrome, which was partially informed by her son’s previous diagnosis with Tourette’s:

Well, I used to be on quite a lot of stuff. They had me on 40mg of Prozac, which wasn’t a lot, but they just cut me back. So that was fine. And, we’re just staying on the maintenance dose. And I’m on Clonazepam to help me sleep and to also watch the movements and stuff. And I used to be on 2mgs, so they’re just trying to figure that out because my psychiatrist thought I had Tourette’s so she had me on some stuff for Tourette’s as well too. So anyway, she’s also got me on the Haldol. Yeah. She had a bunch of the stuff she had given me as well as the Prozac. So that Ativan. She had the Ativan, and then I went on the Clonazepam, on top of all of that other stuff.

This diagnosis influenced Grace’s treatment until her psychiatrist began to suspect she was experiencing the symptoms of another illness. Although there was no history of HD in Grace’s family (her affected father had also been misdiagnosed with Obsessive-Compulsive Disorder), during the differential diagnosis process, one of the diseases her psychiatrist eventually thought to rule out was HD, leading to Grace’s eventual genetic testing for the disease.

Grace’s psychiatrist remained cognizant of her condition and eventually reached the correct diagnosis. It is noteworthy that Grace, unlike many of the other individuals whose misdiagnoses have been recounted in this report, did have the beginnings of the characteristic chorea of HD. Indeed, it was often the appearance of this chorea that led psychiatrists to suspect and then revise their original mental disorder diagnosis. Given that Grace was misdiagnosed while expressing a symptom that is associated with neurological damage, as well as more specifically with HD, it is quite surprising that she was still misdiagnosed, especially given the low prevalence of adult onset Tourette Syndrome (APA, 2000). However, this case does
highlights the difficulty a clinician might have in dropping their epistemological lens to perceive the situation from the vantage of another.

**Genres and Calculation**

This chapter has highlighted the disjuncture between psychiatry and neurology, as described by members of both communities (Cunningham et al., 2006; Price et al., 2000; Schon et al., 2006a; 2006b; Selwa et al., 2006). The ramifications and tangible consequences of this epistemological divide have also been demonstrated through the experiences and recollections of those in the HD community. Although the problems of misdiagnosis and potential solutions have been forwarded in the numerous psychiatric case studies discussed above, these solutions have clearly not led to a substantial amelioration of these misdiagnoses. It is at this juncture that social theory, specifically the works of Latour and Bakhtin, can provide helpful insights by elucidating issues regarding the separation, activation and translation of psychiatric epistemology. First, Latour's concept of oligopticons will be used to further trace the disjuncture between neurology and psychiatry. Second, both Bakhtin's notion of speech genres and Latour's concept of metrology will be used to describe problems with psychiatry's operation, as well as to suggest a possible remedy for this epistemological disjuncture. Crucially, this perspective provides a contrast to the prevailing view in social sciences literature that psychiatry is becoming neurologically dominated.

Neurology and psychiatry both focus their interest on the same organ of the human body, the brain; however, whilst they observe the same object, they do so in different ways. Here, Latour's concept of oligopticons provides us with an avenue to further explore the connections and separations between the two disciplines. Latour (1987; 2005) describes oligopticons as the numerous centers of observation and calculation operating within society. In contrast to Foucault's (1975) concept of the panopticon, which exerted a complete and unobscured gaze,
oligopticons are less comprehensive, but, as Latour (2005) notes, “what they see, they see it well” (p.181). Using this concept, we can understand neurology and psychiatry as constituting two distinct oligopticons, each observing, describing and operating in relation to the brain. Latour’s concept would suggest that, even though these disciplines do observe aspects of the brain in great detail, neither of them perceives it in its totality, with neurology focusing on alterations and injury to the organic matter of the brain, in contrast to psychiatry’s observation of changes in mood, personality and mental disorders.

These boundaries can be traced in greater detail by a brief comparison of Huntington’s Disease (HD) to Alzheimer’s Disease (AD). Both of these diseases are defined as neurological, but in contrast to HD, the DSM-IV entry for AD is a formal diagnostic category (APA, 2000). The entry for AD reviews all the major features of the disease, which are distinctly psychiatric and cognitive in nature. Thus, although the disease is caused by neurological damage (like HD), its effects are completely psychiatric/cognitive (unlike HD). Accordingly, even though AD, like HD, is a neurological condition with psychiatric/cognitive effects, it does not exhibit the same disjuncture between neurology and psychiatry, as both of these disciplines can account for its features. This disciplinary overlap does not occur with HD, perhaps as a result of the motor ability deterioration caused by the disease; however, these motor symptoms do not explain the omission of HD’s numerous psychiatric features from the DSM-IV or its sole inclusion as a subtype of dementia. Thus, we have two diseases, both neurological and with severe psychiatric symptoms, but with only one being accessible to psychiatric observation exemplifying the problematic and somewhat arbitrary boundaries between these two disciplines.

While Latour’s concept of oligopticons provides us with a better understanding of the related but disconnected relationship between neurology and psychiatry, Bakhtin’s (1986) notion of speech genres allows us to better connect psychiatric observations to their effects on patients by focusing on the discursive nature of the misdiagnosis. The act of diagnosis employs
specialized language and forms of communication specific to a given medical institution, as demonstrated by psychiatrists’ misdiagnosis of HD as a psychiatric disorder. These variations of the diagnosis’ context and style represent what Bakhtin (1986) called a speech genre. Although there are as many forms of genres as there are types of communication, each genre has its own style, places its own demands, and commands a specific type of knowledge to be utilized. Those individuals who are fluent and communicative in one genre can be silenced when confronted with another:

Many people who have an excellent command of a language often feel quite helpless in certain spheres of communication precisely because they do not have a practical command of the generic forms used in the given sphere... Here it is not a matter of impoverished vocabulary or style, taken abstractly: this is entirely a matter of the inability to command a repertoire of genres of social conversation (Bakhtin, 1986, p. 80).

Importantly, Bakhtin (1986) notes that the inability to converse in a given genre is often the result of discomfort and unfamiliarity, rather than lack of intellect. This appears to be the case in HD misdiagnosis. It is not that the psychiatrists making these misdiagnoses are inept, but rather are framing the symptoms of an individual with HD through a psychiatric, rather than a neurological or neuropsychiatric, diagnostic genre. What this chapter’s participants, as well as the previously discussed psychiatric case studies, exemplify is more substantial than just an awkward distance between the epistemologies of two different disciplines that both happen to study the brain. Rather, these experiences illustrate how these epistemologies are enacted and applied to individuals, and how misdiagnosis is subsequently translated to inaccurate treatment. Therefore, what has thus far been described as a disjuncture, or “great divide”, between two disciplines can be stated more accurately to be a diagnostic problem of genres. A critical difference between the latter and the former is that, regarding the latter, Bakhtin (1973) provides a means to ground this problems of epistemologies in the actions of specific individuals by explicitly connecting speech to actual utterances (i.e., misdiagnoses). In contrast, the psychiatric
and neurological commentaries focus on abstract issues of pedagogy, rather than discourse and specific utterances. Bakhtin further emphasizes that utterances are dialogical in nature, connecting the speaker to the addressee. As such, we are directed to the dialogical utterance of the misdiagnosis as the event that actually connects epistemology to everyday interactions.

In this context, it is specifically the utterances elicited within the context of HD misdiagnosis that place psychiatrists into a diagnostic problem of genres. Working within the genre of psychiatric diagnosis, it is unsurprising that an individual presenting with both a familial and biographical history of Major Depressive Disorder is diagnosed with Major Depressive Disorder (e.g., Duesterhus et al., 2004), or that an individual, such as Grace, who appears to have problems with anxiety and twitches is diagnosed with Tourette Syndrome rather than HD. The diagnostic problem of genres faced by psychiatrists is further compounded by the inadequate presentation of HD within the DSM-IV. The fact that it is neurologists rather than psychiatrists who diagnose HD is obviously related to this omission. However, even though psychiatrists are not be able to diagnose HD, ample evidence has been presented that they can misdiagnose and subsequently mismanage the treatment of individuals with HD (Duesterhus et al., 2004; Jardri et al., 2007; Stewart, 1987; Tost et al. 2004). The DSM-IV also fails to offer any suggestions or warnings to psychiatrists through the means of differential diagnosis that they might need to approach individuals who seemingly present with psychiatric problems from an alternative genre.

It is this omission from the DSM-IV that returns us to the work of Bruno Latour (1987; 2005), specifically his discussion of metrology. Neurology and psychiatry are related and often harmonious disciplines, but an ambiguous disease such as HD accentuates their lack of a common language, or shared speech genre. Stated in Latour’s terms, these two disciplines lack a system of metrology, which he defines as a system of standards that allows observations made at one location to be interpreted at another:
Since without standards like the watt, the Newton, the ohm, the ampere, that is, without the Systeme International d'Unites, there would be no global of any sort because no locus would have the 'same' time, the 'same' distance, the 'same' weight... All sites would be incommensurable for good (2005, p.228).

Once more, although neurology and psychiatry share the same organ of interest, they describe and observe it in different terms, with different aims and in different units. Thus, whilst one group of professionals seeks to uncover damage to the structures, the other seeks to identify changes in behavior patterns. When encountering an illness of complex and ambiguous properties, such as HD, the fissures between these two perspectives on the brain become activated. In lacking a metrology, or a system of standards, there is no indication for clinicians encountering a set of ambiguous symptoms on how to proceed, signaling that they might have to convert their observations into those of a related discipline.

The absence of a discussion in the DSM-IV of the risk and history of misdiagnosis of HD as a psychiatric disorder thus takes on new significance. The DSM-IV, by providing uniform definitions and descriptions of mental disorders, acts as North American psychiatry's metrological artifact. It is precisely within such a text that guidelines should be provided for clinicians for when they might need to use a different system of observation, or diagnostic genre.

Curiously, while the DSM-IV omits this information, the International Classification of Disease (ICD), published by the World Health Organization (2007) and used by European clinicians, does include an entry of HD as a neurological condition, in addition to its entry as a precursor for dementia (in consecutive sections no less). Although a more comprehensive diagnostic tool will not entirely eliminate the risk of misdiagnosis, having this information available, as in the ICD-10, does provide the clinician with alternative interpretations of their patient's symptoms.

By focusing on observation and language, this analysis highlights the disjunctures between psychiatry and neurology. This outcome is particularly important given the predominant
evaluations of psychiatry in social science literature describing the discipline as increasingly dominated by neurology. Two exemplars of this perspective are Rose’s (2007) *The Politics of Life Itself* and Whitaker’s (2002) award-winning *Mad in America*. Whitaker documents the psychiatric adoption of neurochemical explanations of schizophrenia, which, in presenting the disease as a neurochemical imbalance, justified the sale of pharmaceuticals to adjust this imbalance. Similarly, Rose draws on a number of psychiatric accounts to emphasize the discipline’s adoption of neurological modes of explanation. He includes numerous quotes from psychiatrists who perceive the application of neurotechnologies as an end to distinctions between mind and brain, as well as the organic (neurological) and functional (psychiatric) divisions of the organ. Like Whitaker, Rose also connects the neurological movement of psychiatry to the sale of pharmaceuticals.

These compositions of psychiatry highlight a number of important issues, not least of which is the troubling association between the discipline and pharmaceutical companies. However, their connections between psychiatry and neurology omit a number of critical points that can only be illuminated by studying misdiagnosis. For instance, the psychiatrists that Rose quotes are contemporaneous with the authors of the previously discussed misdiagnosis case studies (e.g., Duesterhus et al., 2004; Jardri et al., 2007). Rose’s psychiatrists extol the eradication of the mind/brain distinction while, in contrast, the case study authors highlight very tangible gaps between neurology and psychiatry, gaps also noted by a number of psychiatrists and neurologists (e.g., Price et al., 2000).

Critically, a psychiatrist’s ability to draw on neurological information does not change the types of diagnoses that he or she has available. Rather, it provides an alternative mode of explanation for these pathologies. Similarly, this information does not prevent the misdiagnosis of neurological conditions, but instead ironically justifies these misdiagnoses with neurological language, tools and data. To put it succinctly, a clinician’s sensitivity to neurochemicals does not
prevent them from diagnosing a person with HD as schizophrenic, it merely changes the
evidence drawn upon to reach the conclusion, which, while being more neurological, remains
just as inaccurate.

Accordingly, by focusing on language and observation, we can understand the
interactions between psychiatry and neurology on a more minute level. Approaching their
relationship in this way focuses our attention on the interpretation of borderline and ambiguous
cases, such as HD. Such an approach highlights the numerous discords between neurology and
psychiatry omitted in larger, more systemic analyses, such as those forwarded by Rose (2007)
and Whitaker (2002).

Conclusion
Psychiatry has been externally critiqued from a number of vantage points, with researchers
suggesting that the diagnostic ability of psychiatry is inaccurate (Brown, 1990; Good, 1993;
Rosenhan, 1977), distorted by larger social scripts (Floyd, 1997; Smith, 1975; 1978; 1983) and
insensitive to social factors related to mental illness (Jarvis, 2007; Sayre, 2000). The discipline
has also been questioned internally, particularly in regards to its relationship with neurology
(e.g., Cunningham et al., 2006; Price et al., 2000). However, the majority of these commentaries
focus on either the epistemological structure of psychiatry or the nuances of clinician-patient
interactions.

Engaging with the work of both Latour and Bakhtin, this chapter has sought to unify
these two perspectives by highlighting the epistemological disjuncture between psychiatry and
neurology, as well as its actual consequences for living individuals. Whilst these consequences
have been suggested through the numerous case studies on misdiagnosis (e.g., Duesterhus et al.,
2004; Jardri et al., 2007) this report has added nuance to this dialogue by offering the accounts of
the individuals living with HD. Their experiences underscore the pain, confusion and grief that
can result from misdiagnoses, factors that are not made particularly salient in either case studies or conversations on epistemology.

With an accurate genetic screening tool available for HD, there would appear to be few reasons for misdiagnosis. However, before a genetic test for HD can be performed, the clinician engaging the patient must consider HD as a possibility. Given the recent reports on the progression of HD (Duff et al., 2007; Marshall et al., 2007; Stout et al., 2007), there is a substantial likelihood that the clinician might be a psychiatrist. However, the omission of important information on HD within the DSM-IV, as well as the “great divide” between neurology and psychiatry (Price et al., 2000), situate these clinicians at a disadvantage and accordingly place their patients at-risk of misdiagnosis.

This report’s participants, and the neuropsychiatric features of HD, highlight some of the problems with multiple, discordant, healthcare epistemologies. However, HD also offers a forum and opportunity to make progress on addressing such disjunctures. For instance, although the psychiatric case studies on HD misdiagnosis shared a common mistake, they also shared common recommendations: the need for better understanding between psychiatry and neurology (Appollonio et al., 1997; Duesterhus et al., 2004; Jardri et al., 2007; Stewart, 1987; Tost et al., 2004; Yu. 2004). These statements were echoed in the numerous commentaries on the division between psychiatry and neurology, which stated the need for better education to achieve this understanding (Cunningham et al., 2006; Price et al., 2000; Schon et al., 2006a; 2006b; Selwa et al., 2006).

Drawing on the work of both Latour and Bakhtin, this report suggests that establishing such an understanding requires more than reforming the education of medical students on this topic. A successful remedy to this disjuncture also needs to acknowledge that misdiagnosis is the instance where such epistemological divides are operationalized and are translated into very real effects for individuals. Accordingly, a pivotal step in decreasing the chances of the misdiagnosis
of HD, as well as similar misdiagnoses, would be the implementation of a more comprehensive diagnostic tool. Such a tool would give clinicians an opportunity to translate their observations into the genre of alternative epistemologies, as well as providing a sensitization to instances where such translations are necessary. Although a more pluralistic diagnostic guide will not eliminate misdiagnosis, or address many of the other external criticisms of psychiatry, it may affect the way that its epistemology is enacted, which, as this report suggests, should have tangible benefits for the individuals encountering it.

Now that the difficulties caused by the psychiatric aspects of HD have been reviewed, this thesis will now address other health services. Accordingly, the next chapter will review participants’ evaluations and recommendations of physiological and psychosocial services, as well as disability regulations.
CHAPTER 3: HEALTH SERVICES AND BUREAUCRACIES

Introduction

Huntington Disease produces a diverse set of symptoms. Individuals may experience cognitive abilities, psychiatric disturbances and loss of motor function (Paulsen, 2004). People with HD may also experience these symptoms in markedly different ways. Accordingly, individuals with HD have multiple and nuanced healthcare needs (e.g., Skirton, 2005). Health professionals, such as those at the Huntington Disease Medical Centre (HDMC), attempt to address these diverse needs by providing a multi-disciplinary team of neurologists, geneticists, genetic counselors, researchers, social workers and psychiatrists (Centre for Molecular Medicine and Therapeutics, 2009). However, people with HD also encounter numerous other healthcare workers and bureaucrats external to specialized clinics, thus interacting with both individualized tailored care and generalized public healthcare services.

These patient-practitioner interactions are a frequent focus of social health research (e.g., Foucault, 1973; Mol, 2002; Rose, 2007), which has provided a number of important suggestions for better care delivery. For example, Kleinman’s (1978) helpful delineation between the numerous perspectives on illness provides a means for better communication between patients and practitioners. Likewise, Frank’s (1991; 1995; 2004) work has also provided a number of helpful recommendations, such as highlighting the need for dialogical and reciprocal understanding between patients and providers, what he calls a renewal of generosity.

Recent reports on HD have suggested that the preponderance of accounts of the disease are written by either medical researchers or clinicians (e.g., Wexler, 2008), with a number of researchers critiquing the exclusion of individuals with HD from research that focused on their care (Gibson, Timlin, Curran & Wattis, 2004; Nygaard, 2006; Roger, 2007). This is an important omission both with regards to the unique healthcare interactions of individuals with HD, as well
as the numerous benefits that can be introduced by involving the perspectives of the ill on health services (e.g., Frank, 1995; 2004; Kleinman, 1978). This omission is additionally compounded by the pervasive concern that Canadians have vis-à-vis the tenability and quality of their healthcare (Soroka, 2007).

This chapter engages with these issues by focusing on the encounters of individuals with HD with both medical services and health bureaucracies. The analyses will discuss the perspectives of individuals with HD on four discrete sites of healthcare: services provided by the HDMC and the HDRC; peer support offered by the HSC and HDRC; services provided by general health professions (i.e. those who are not HD experts); and the regulation of disability.

**Previous Discussions of Huntington Disease Care**

Reports on HD have made numerous health services recommendations. Foremost amongst these are the protocols for predictive genetic testing (Benjamin et al., 1994). Benjamin and colleagues outline a number of recommendations and considerations for the ethical and safe delivery of the genetic test for HD. They suggest numerous counseling sessions for those desiring to take the test, as well as screening to assess suicide risk. Their report also suggests that healthcare professionals may wish to delay formal diagnostic procedures for individuals who have received a confirmatory test until the individuals have had a reasonable amount of time to process their genetic status. Lastly, the protocols, as reflected in both their content and authorship, emphasize interdisciplinary cooperation in health provision for individuals with HD and their families.

There have been a number of similar recommendations around the provision of genetic testing. Among Cox’s (1999) suggestions was that individuals receiving genetic testing should be given more autonomy, specifically in terms of directing the content of counseling sessions. She also advised counselors to avoid the use of terms such as “positive” and “negative”, as well as minimizing focus on test outcome. Bombard (2008) also provided feedback for genetic
counselors, based on her examination of HD and genetic discrimination. Foremost, she noted that HD centres across Canada were not consistently informing their clients about issues relating to genetic discrimination. She also advised health professionals that individuals receiving a negative genetic test, as well as those who refrain from testing, also reported experiencing genetic discrimination.

Other researchers have also echoed Benjamin and colleagues statements on the importance of integrated and multi-disciplinary care for people with HD. McGarva (2001) noted that, due to the broad range of potential problems that individuals with HD and their families may encounter, health professionals should actively solicit the input of their peers. Likewise, Skirton and Glendinning (1997) encouraged nurses to offer patients and their families access to psychiatric services, neurological services and nutritionists. Skirton (2005) also emphasized the importance of including nurses in the management of HD. Nance (2007) made a similar argument in her discussion of what she calls the “Huntington disease molecule”, wherein the patient occupies the conceptual centre, surrounded by their loved ones, and then connected to a team of health professionals in a secondary ring. Nance suggests that this team should be led by a physician and include rehabilitation therapists, psychologists, genetic counselors, and social workers. Lastly, given the high numbers of individuals who may be involved in the care of an individual with HD, Kent (2004) emphasized the importance of coordinating such large teams, though not necessarily by a physician.

Finally, as reviewed in Chapter 1, there have been numerous recommendations on issues relating to death and palliation. Dawson (2004) recommended that end of life care be tailored to the needs of the specific individual, and stated that providing in home support was particularly beneficial. King (2005) reviewed the medications that she used to treat an individual dying from JHD, whilst also noting the ethical dilemmas she encountered during treatment. Several researchers have also discussed issues of suicide, with studies cautioning health professionals in
relation to suicide risk and genetic testing (Farrar, 1986; Paulsen, 2001). The most recent study by Paulsen (2005) has further suggested that health professionals need to be aware of suicide throughout the client’s experience of HD, particularly when they begin to experience notable symptoms and disability.

The research community has provided a litany of suggestions to improve care for individuals with HD. Importantly, the majority of these articles highlight the need for health professionals to cooperate and to produce care that is tailored to the needs of their specific client (e.g., Benjamin et al., 1994; Kent, 2004). That said, the bulk of these suggestions focused on one of two issues: hospitalized/clinic care (e.g., Skirton, 2005; Nance, 2007) or genetics (e.g., Bombard, 2008; Cox, 1999). These are exceptionally important issues to consider, but attention must also be given to the area of care between these two points, that is, interactions with medical services after their genetic testing, but before end of life issues develop.

**Huntington Clinic Services**

Every member of the HD cohort, without exception, had a positive attitude towards the health professionals at the HDRC and the HDMC. Indeed, no answers were generated for the questions (see Appendix 4) that requested suggestions for a new service for people with HD, or identification of an area of their disease that did not receive adequate support. In the author’s previous experience as a health researcher, as well as Canadians’ overall evaluations of healthcare (Soroka, 2009), such an attitude is rare.

Many participants spoke about the services they received from the University of British Columbia (UBC; where the HDRC and HDMC are located) in very general terms. While these individuals could not pinpoint anything specific about the HD team that led to their positive evaluation, they did note that “everything was good”, or that the team was “just fantastic”. Furthermore, although the HDMC and HDRC were a considerable distance away from numerous
participants, no respondent reported being bothered by the distance they were required to travel. Indeed, several participants described how they turned their visits to UBC into an excuse to visit family, or to have a brief vacation in Vancouver.

Individuals who were able to describe their attitudes in greater detail always connected their positive experience to the staff at UBC. Participants related numerous stories of how members of the HD team supported them during particularly trying times. Others noted that the staff were always happy to see them, and made themselves available outside of normal work hours. Delores (34, gene-positive), discussed the empathy of the staff at the HDRC and HDMC:

I talk to [social worker] more about my dad. I’m in touch with them. Like they check in and make sure I’m doing okay and if I need them or if I’m having trouble I know I could phone or email and they’d have me in there in a heartbeat. I feel supported, very supported. I feel very connected as well. I’ve never seen anything like it. And – where it’s a direct comparison is at the – within like months of each other [husband’s] dad got diagnosed with Parkinson’s. And so that we’ve been at the UBC Parkinson’s clinic, but you go there, you see a doctor, you go home. There’s nothing. And like [mother-in-law] knows about the HD clinic and she’s like “well how come there’s nothing for Parkinson’s? Like how come there’s nothing? Like no support” It’s very unique. I’m not aware of anything else like it. Where you have like a doctor, a social worker, a psychiatrist if you need it, like everybody is at your disposal!

Delores connects the availability of the health professionals, as well as their communication skills, as helping her feel “supported”, thereby highlighting their interpersonal abilities as essential to her care. She also makes an explicit comparison between the HD clinic and the UBC clinic for Parkinson’s Disease, which is also a neurological illness that affects movement. Here Delores describes how important the interdisciplinary team at the HD clinic has been for her, as she is able to access a diverse set of professionals. In contrast, her mother-in-law states that she has “nothing” at the Parkinson’s clinic. Interestingly, while the Parkinson’s clinic does provide access to a doctor, this account suggests that the psychosocial support is also an important factor. That is, while Delores’ mother-in-law is receiving biomedical information, she feels comparably underserved without the additional network that Delores has access to.
Other participants made similar positive connections between strong interpersonal skills in tandem with comprehensive health services. For instance, Samuel (47, diagnosed in 2003), said,

They make sure my medications are okay. And they’re, great researchers too and just good people overall you know? They’re very caring people and they understand, you know? It takes a lot to look at a person with Huntington’s so, in their later stages. So I’m sure it’s very difficult to do.

Samuel notes that the UBC staff are competent medical professionals and researchers; however, equally important to him is that they are caring individuals who are willing to do what he perceives is the difficult work of personally engaging with HD.

The importance of an empathic medical staff was also related by Cecilia (42, gene-positive), as she described a recent encounter with one of the mental health personnel at the HDMC:

He [HDMC staff member] doesn’t just deal with the person that is affected with it, he deals with the whole family. He’s just as concerned with the partner or the daughter or the husband or whatever as he is for that person. And he spends just as much time talking with them about where they are at, and not just with dealing with HD, but in their whole life. He’s just very thorough. He’s on the ball. This guy is just amazing. You know, its just like man, “how can anyone be that smart and that relatable?” [laughs]. And he just has this way, and my dad is a very difficult person, and he was a very difficult person before he even had this, and this man can relate to him and its like, “wow! I don’t know how you do it!” So he’s just a very special and brilliant person.

Once again, Cecilia cites the interpersonal skills of a UBC professional as being central to her positive review of the services she receives. She also underscores the importance of engaging with the entire family, complimenting the inclusion of her unaffected partner in her care. Furthermore, as well as being gene-positive herself, she is also a caregiver for her father and has been similarly integrated with his care.

Both the HDMC and the HDRC received universal praise from participants. Individuals described the benefits accrued from both treating HD as a family disease, as well as providing
interdisciplinary care. Viewing the disease in such a manner has been advocated in both academic literature and protocols for HD predictive testing (Benjamin et al., 1994); the benefits of these recommendations are quite notable. Participants also stated that the engagement and empathy they received from staff members was particularly important, suggesting that being engaged as an individual was as important as the services.

**Peer Support**

There are numerous forms of psychosocial support available for individuals with HD. These services predominately involve one-on-one interactions with a health professional. However, the HSC, through its HDRC, provides two additional forms of group-based psychosocial support. While individuals benefited from these resources, they also provided suggestions to increase their applicability.

The first form of psychosocial support was an annual camp, which takes place over a week in a small mountain town near Vancouver. This camp provides individuals with mild to moderate HD an opportunity to engage with similar individuals, and in a number of therapeutic and physical activities. The camp is a popular event and has a waiting list of participants (S. Tolley, personal communication, June 2nd, 2009).

The second source of psychosocial support are the support groups operated in both the Greater Vancouver area and Victoria. The Vancouver support group is located in a central suburb and meets bi-weekly. This group includes both individuals with HD and caregivers, and is coordinated by a social worker. The Victoria support group meets thrice monthly. The first meeting is primarily social and is typically a potluck lunch. The second and third meetings are exclusively for people with HD and caregivers, respectively.

Numerous respondents within the HD cohort had been attending one of these peer supports for years. Individuals were quick to note the benefits of such support, even those
participants who claimed that they “rarely talked” or “just listened”. Kenneth (76, diagnosed in 2003), reflected on his interest in engaging with these events. He said,

Yeah, that’s a fabulous experience. A five-day experience up at [mountain town], but you had to have Huntington’s to go. Now, for the first two years after being gene-positive, I didn’t get to go so I was almost begging her [HDRC staff member], “come on, come on, come on! [Both laugh] give me a diagnosis!”

Comically, Kenneth was hoping to receive confirmation of symptom onset so that he would be eligible to attend the camp. However, his joke communicates his intense interest to be able to participate in such activities. This is particularly noteworthy as older males are often identified as not wanting to participate in psychosocial support groups (e.g., Mo, Malik & Coulson, 2008; Oliver, Pearson, Coe & Gunnell, 2005).

Participants at these groups reported receiving numerous benefits from attending. Often it provided them a forum to discuss issues of importance, while other times, it provided one of their few social outlets. For instance, Gerald (52, diagnosed in 1996) lived in a very isolated area, meaning that being able to attend the camp provided a rare occasion for socializing. Similarly, Becky (36, diagnosed in 2007) was unable to leave her home without the aid of her husband, who worked during the day, and, like Gerald, was quite isolated outside of her experience at camp. Louis (64, diagnosed in 2004), described the support group as a similarly social affair:

We have a Huntington’s support group and it meets tomorrow night. The first week is a family get-together and everybody brings food and we eat. And then next week is just for me [people with HD]. And this is with a social worker. And then the week after that the spouses have a meeting with her. So that if they have a problem or want to discuss anything they can. I look at some of the other people and I don’t think that I’m as bad and that I hopefully don’t reach that stage, and that’s good.

While describing the social features of the group, Louis also notes two other benefits. First, the group that he attends offers support to both individuals with HD and their caregivers. Second, the group provides him with a way to put his own symptoms in context; by seeing individuals dealing with worse symptoms, he feels better about his own symptomology. Accordingly,
individuals who attended these group supports derived benefits both from the opportunity to socialize, as well as learning more about their disease.

There were also several participants who did not attend these group supports. One of the primary reasons that individuals offered for not engaging with these services was anxiety. Although he eventually attended the support group, Keith (55, diagnosed in 2003), was initially inhibited by such anxiety. He said,

Well, I mean, it was kind of weird. I mean, a bit nerve wracking, because I've sort of always had a performance anxiety, or social anxiety. But I mean, if I can't open up there [support group] where can I? ... So the first time, it was a bit nervy, but since then I've been fine. So yeah it was fantastic in retrospect.

Keith now regularly attends the support group in Vancouver and he notes that it has been a “fantastic” experience for him, and an opportunity to disclose. However, it is important to note that his social anxiety, as well as his initial fear of disclosure, initially prevented him from attending the activity he now enjoys. Although Keith overcame his shyness, it still prevented other individuals from attending the support group. For instance, Samuel (47, diagnosed in 2003) stated, “I've had lots of opportunities to go to camps and things out here but, that's about it. I'm just a bit shy to go to camp and those things.”

Several individuals did not consume these services because they were shy or wary about disclosure. However, amongst individuals who were experiencing low-level symptoms, there was a feeling that the current support groups were not applicable to their unique circumstances. For instance, Cecilia (42, gene-positive) said,

I don't, at this stage, feel that I need to. I feel that I get, as far as support for me goes, I get a lot from friends and family and as far as support for how to help my dad, just with my family and the clinic and all the research and material I have, I just, at this stage, don't feel that I need it.

Cecilia believes that she is currently receiving enough support from her social circle. Indeed, when discussing support in the above quote, she identifies herself as much as a caregiver for her
father as someone living with HD. Critically, she emphasizes her “stage” several times, thus suggesting that, as an asymptomatic and presumably healthy individual, such support does not apply to her.

Although Phyllis (23, diagnosed in 2008) had received a diagnosis of HD, this only occurred a few months before our interview. Like Cecilia, she did not attend any form of peer support. However, she was markedly more explicit about her reasons. She stated,

Whenever I even see someone who looks slightly like they might have Huntington’s Disease or anything like that, it just makes me freak out and I don’t want to see it ‘cause I don’t like seeing people like that. So I find with all the support groups, it’s like other people whose like families or who have Huntington’s Disease and who might be progressed further than me. And I don’t want to have to be around them in a room and have to see them. It would be just too much for me right now, so that’s also why I’m not a part of anything like that.

Phyllis expresses a palpable fear of individuals with HD, so much so that she is averse to seeing people who appear symptomatic. Witnessing people with HD is almost a reminder of what she might be facing in the future. As such, in contrast to Louis’ statement above, being around those with more severe symptoms is not therapeutic, but almost traumatic. When asked if she would attend a form of support that was based around individuals with fewer symptoms, she responded, “yeah then I would go to that kind of thing, yeah. I just can’t stand to see the sight of it [HD].”

These feelings of aversion and the need for stage-specific support were perhaps best described by Delores (34, gene-positive). She participated heavily in HD events and volunteered for the HSC. However, like her sister, she did not attend the peer support group:

There should be – and I’ve said to [social worker] too – like an HD positive support group. I really think so. And also I think it’s not necessarily that healthy for people who don’t have it yet to sit with the people who do. I just think instead of walking away thinking “oh I feel better” you’d think “oh god, look what I have to look forward to”. Like if you had to sit there like in a group of people and you were like in the far reaches of Huntington’s or halfway through the stages of the disease and you knew you were inheriting the gene you don’t walk away thinking, “I feel so much better” you know? It’s different if you’re sitting around with a bunch of people who are gene positive and you’re all in the same boat and you’re presymptomatic and, you’re trying to relate to people right, and you relate to the person that’s not symptomatic.
Delores suggests that individuals without symptoms have more in common with one another and would thereby provide a better network for communication and support. Like Phyllis, she also expresses apprehension about attending a support group with symptomatic individuals. Indeed, she questions whether it is even psychologically healthy for people without symptoms to witness what they may experience in the future. She continued by also questioning whether it was appropriate for symptomatic individuals to be around the asymptomatic:

And maybe vice versa. You know what I mean? I don’t know, it’s like seeing what you don’t have. Like seeing what you’ve lost. I don’t know. Maybe it wouldn’t bother them [symptomatic members of a support group, I don’t know. I just think if you had symptoms and you were struggling and you were struggling with chorea and struggling with all this stuff and you’re in a support group with people who have no symptoms and are perfectly fine you’re gonna sit there like, “why are you here? Why do you need support, there’s nothing wrong with you”. You’re gonna relate better to the person that says, “hey, you know when I picked up my coffee mug this morning I spilt it all over myself”. “Yeah, you know, that happened to me too”. Like you know? It’s just like – it’s a completely different thing. We’re all in the same boat, but it’s just nice to talk to other people who like have the same sort of circumstances you know? ‘Cause it’s a unique circumstance. It makes you feel less isolated, you know like you can relate – it’s nice to relate to people.

Here Delores suggests that an integrated support group is also unfair for the symptomatic. Not only must they encounter what they no longer have, but they will also have a problem relating to individuals without their specific concerns. Delores further hypothesizes that individuals with symptoms will perceive the asymptomatic as illegitimate consumers of peer support. Despite these numerous reasons for not wanting to attend current support groups, Delores still states her desire to find similar people to relate to. While participating in the general support group may be difficult, she suspects that dealing with individuals without symptoms would help her feel less “isolated”, which is one of the primary benefits outlined by those who did attend the groups.

Perhaps Delores’ most substantial suggestion is that being symptomatic is a “completely different thing” then being asymptomatic. This attitude is echoed in the statements of both Cecilia and Phyllis, who perceive current support services as not applicable or outright
intimidating in their current situation. Although the individuals who did consume these services emphasized their enjoyment of them, those with comparably less symptoms did not feel comfortable engaging in such groups.

General Medical Services

While participants levied universal acclaim on the staff at the HDMC and HDRC, other health professionals were not as well perceived. The majority of respondents had at least one instance where they encountered a health professional who was ignorant to HD. This ignorance had both psychological and physiological consequences.

For instance, several participants recalled negative interactions with health professionals when offering them information on their disease. When diagnosed, individuals would bring a pamphlet explaining the diagnosis to their general practitioner (GP). This pamphlet explained HD from the physician's perspective, while also providing a list of drugs that individuals with the disease should not consume. Several participants recalled that their GP was particularly reticent or even combative about receiving such information. For instance, Keith (55, diagnosed in 2003), stated that his doctor told him that he “wasn’t going to read that” and that he didn’t “get paid” to review such information.

Similarly, respondents also noted that their GPs often had difficulty talking to them about their disease. Instead of consulting the HDMC or asking the patient about HD, the doctors would frequently take their own course of action. As Kenneth (76, diagnosed in 2003) stated,

I think that they need to listen more to their patients because with a disease like Huntington’s the person is usually able to keep his facilities, his mental facilities, they don’t stop thinking. They don’t stop being creative and that. If the doctors would only ask the patient oftentimes the patient knows more than the doctor does about the Huntington’s and that would be a big step forward.
Kenneth suggests that HD patients are perhaps being ignored due to an inaccurate view of their mental capacities, although the patients may actually have more knowledge of HD that their doctor. The communication dynamic highlighted here by Kenneth, and above by Keith, provides an interesting contrast to the communication skills displayed by the staff at the HDMC and HDRC, which was central to their positive evaluation by respondents.

The reluctance to learn about HD was typically a problem that individuals encountered with their GPs. However, health professionals who were also experts in genetics or diseases similar to HD also expressed ignorance related to the disease. Donna (47, diagnosed in 1999) described one such encounter with a medical geneticist:

I know once from Dr. [name], he told me that HD goes back to the days, over in Europe, when sheep farmers had sex with sheep when they were out for a while. They figured that’s where it came from. And so some people think HD is dirty and something to be kept secret.

Although the doctor mentioned above actually conducted tests for HD, he also disseminates a false explanation of its origins that is similar to stigmatizing explanations of HIV/AIDS (Sabatier, 1988). These proposed origins are both offensive and shaming to individuals with HD, suggesting that they inherited the disease from relatives who engaged in bestiality. Donna further connected the shame that individuals feel from such comments as partial motivation for why some people with HD contemplate suicide (see Chapter 1). As such, the ignorance that some health professionals have around HD can be translated into very tangible, and harmful, consequences.

Health professionals’ reluctance to learn about HD typically caused strained relationships, communication breakdowns, or a change in doctor. As suggested by Donna, another possible consequence of this ignorance was psychological harm. However, there were also several instances where a health professionals’ neglect for HD also resulted in physiological harm. Specifically, several respondents stated that they had been given incorrect medications by
a health professional. One respondent tearfully described how her sister had very suddenly
become near catatonic after taking a new medication, but she could not convince her physician to
contact the HDMC for advice. Likewise, Cecilia (42, gene-positive) described how her father’s
GP had prescribed him a medication that was not safe for individuals with HD. She said,

My dad’s old family doctor prescribed a drug that is incorrect, that should never be
prescribed to an HD person. And it was prescribed to him and he did take it and the
effects were awful and I am just so thankful that they were not long lasting. Yeah it was
really bad. We talked about malpractice. Because he was given a list of medications that
if my dad couldn’t get into the HD clinic that he should follow this regiment. “If this drug
doesn’t work, try this drug, if this doesn’t work, try this” type of thing. So he had that
information and he chose not to use it and he chose to prescribe him something that not
only was not on that list but was known that you cannot prescribe that drug to an HD
patient. Yeah, it was bad. We had some serious discussions [about malpractice]. Actually,
it’s as long as my dad continues to get better and, if not, we’ll have another discussion on
that [laughs].

Similar to those stories above, this doctor chose to neglect and contradict HD specific
information. Fortunately, the physiological consequences of this decision were reversed, but they
still necessitated the termination of the client-doctor relationship and nearly prompted a lawsuit.

A central factor in these accounts is the fact that the HDMC is a consultation service
only. That is, the health professionals there may give advice to other physicians, but the
physicians are under no obligation to follow that advice. As such, although the HDMC provided
doctors such as Cecilia’s with a list of safe medications, it is the doctor’s prerogative whether to
use that information or not. Thus, while the HD clinic specifically tailors services for their
clients, external health professionals may not adopt this same perspective.

**Regulating Disability**

Due to its degenerative nature, individuals with HD must ultimately encounter issues of
disability. In many cases, the disease necessitates a cessation of paid labor. The loss of income
requires individuals to access disability benefits and subsequently encounter the numerous
bureaucratic entanglements that such an endeavor entails. In addition to dealing with regulated benefits, moving into the category of “disabled” can affect an individual’s sense of self. Issues around disability will thereby be explored in the context of job loss, accessing disability payments, and consequences for autonomy and identity.

For many individuals, symptom onset signaled their loss of autonomy. One area in which this was particularly salient was the termination of paid labour, as the numerous effects of HD make maintaining employment a difficult task. The psychiatric effects of HD can alter an individual’s mood, creating difficulties between them and their co-workers, while the cognitive decline that individuals may encounter can affect their ability to perform once familiar tasks. Likewise, the motor effects of HD can make routine movements difficult. Accordingly, only four participants were still employed at the time of their interview. Two of these individuals were the gene-positive sisters, who were not experiencing any difficulties fulfilling their work obligations. The other two participants, Phyllis (23, diagnosed in 2008) and Albert (57, gene-positive), both encountered HD related difficulties at their jobs. Albert was formerly a truck driver, but a work related accident, combined with his HD symptoms, made that occupation untenable. Instead of retiring, he decided to work as a part-time computer sciences tutor. At the time of our interview, Phyllis was working at a regional restaurant chain and received regular derision from her coworkers about her job performance. Shortly after our interview, she was fired.

Other members of the cohort shared these difficulties. Gerald (52, diagnosed in 1996) stated that he was fired after an “angry outburst on a job site”. Arthur (67, diagnosed in 1996) began to have “a lot of memory problems” at his job and was eventually asked to work from home before his company was bought out and dissolved. Similarly, Samuel (47, diagnosed in 2003) was working as a dentist when his symptoms started to appear. After a year of noticing his comprehension problems, as well as his “slower memory and also that [he] was getting a bit shaky”, the staff at his dental clinic confronted him about his symptoms. Although initially
angry, Samuel subsequently got tested for HD and was confirmed to have symptom onset, leading to his retirement.

As shown, disease symptoms precipitated retirement or job loss in a number of cases. Participants were then required to live off of their pensions, which were supplemented by disability benefits. Although these sources of income helped provide for the majority of individuals, the bureaucracy around disability payments created numerous difficulties. Individuals who had already secured their payments encountered numerous aggravations. In particular, they were required to give constant updates about the status of their disease. As Keith (55, diagnosed in 2003) said,

"It’s always “has your condition improved”. I think even with the forms for short term, it's the things that you know are going to be “no” so you just put “no.” I mean, they shouldn’t be asking you. So I said that one time, I said, you know, “I have HD, I’m not going to be recovering, there’s no magic cure”. So I had sort of a verbal battle with insurance."

Keith uses gallows humour to vocalize his aggravation in being required to annually update his benefit provider as to whether or not his genetic condition is still present. Several respondents, who similarly did not understand why they were required to answer questions that appeared to have no relevance to their illness, echoed his frustration. In response, Grace (53, diagnosed in 2005), decided to photocopy the form that she was required to submit. When she received a request for an update of her illness she submitted a photocopy of the previous year’s information, which was accepted by her provider.

Both Keith and Grace describe the frustrations involved in receiving disability benefits. However, the most notable difficulty was accessing the benefits to begin with. Phyllis, for instance, was having problems maintaining employment due to her symptoms. As a result of these difficulties, she often needed to access employment insurance. These funds failed to provide her with enough money to keep a basic standard of living; however, she was also unable to access disability benefits as she lacked a formal diagnosis:
They couldn’t see anything like in my brain, like that kind of thing at all. And for the movements and that kind of thing, it’s mostly my hands were like jittery. But how you look at things is different and it’s not at the same level as normal people. My symptoms aren’t that bad right now, but we need to get me on to disability. Because the more stress in my life, then the faster the symptoms will progress. So basically I was like trying to get on that [disability] as soon as possible, because working and everything else was like completely really stressful for me. The diagnosis at the time and everything was completely all very true and everything. It was just when we were filling out the forms for the disability and everything that we kind of made it that I was being more affected, because the government, they’re very weird about things and if it’s not enough, if you can’t function and all this other stuff, then they’re not going to give it to you. But it’s so really frustrating right now, seems like there’s like been block after block after block, and living off $610 [employment insurance] a month is not possible.

Phyllis has described a considerable bind. She currently is only accessing either $610 from employment insurance or the amount of she can earn in the service industry. As a result of these financial restraints, she was sharing an 800 square foot apartment with three other tenants.

Phyllis also states that she is no longer able to function at the level of “normal people” due to her cognitive abilities. Indeed, recent research suggests that these are some of the earliest symptoms (e.g., Duff et al., 2007; Marshall et al., 2007; Stout et al., 2007). However, her disability payment is based on a formal diagnosis, which focuses on motor problems and tangible damage to the brain. Consequently, she finds herself needing to exaggerate her symptoms to be able to secure her disability payment and make ends meet.

Similar to Phyllis, Mark (60, diagnosed in 2009) encountered difficulties in accessing disability benefits. However, unlike Phyllis, these difficulties were predominately tied to his identity. Shortly before his diagnosis, Mark had been working a prestigious job overseas. After the global economic collapse of 2008, Mark was left without employment. Little more than a month after the loss of his job, he was also diagnosed with HD. Transitioning from his comparatively glamorous lifestyle to living with HD provided him with numerous challenges. He said,

I thought I would end my days in the Indian Ocean, and now that’s gone. I don’t know which way to go yet, and what to do with the rest of my life. [Caregiver] is coming over tonight and see if we can work out my expenses. But I haven’t even been living here for
very long and all of a sudden there are all of these people telling me what to do. I met with Dr. [name] to talk about this whole thing, about being disabled. I said “I’m not disabled, I’m underemployed!” I keep saying, “I’m just underemployed for god’s sake!” They’re saying “you got a diagnosis for Huntington and you got to prepare yourself and set up things in your life so you live as stress free as possible”. And apparently the less stress, the better quality of your life. I’m just laughing because I’ve got one foot in one world, and one foot in another. To be disabled and living off assistance, that to me is the hardest part of my process. So I haven’t put the paperwork yet [laughs]. Because my brain keeps saying, “once you go down that route it’s a slippery slope”. But how I feel about it is, I don’t want to be disabled. It is a “disability benefit” and I will be disabled.

Although he is facing substantial economic challenges, Mark cannot yet bring himself to apply for disability benefits. Critically, this reluctance focuses on how he identifies himself. By accepting disability benefits, Mark feels that he is also required to accept that he is “disabled”. This label is additionally problematic as he is still considering resuming paid employment, which is also the only way he can reconnect with his girlfriend, who has not been given permission to visit Canada. Disability benefits are at the nexus of Mark’s predicament, where he must balance the goals of his life with the realities of his illness. In this case, by accepting the label that comes with the benefits, Mark feels he also must abandon his previous life and begin to live as though he were disabled.

These accounts emphasize the numerous difficulties involved with consuming disability benefits. In addition to the lost autonomy of paid employment, individuals must struggle with the label of “disability”. While the funds themselves provided financially for the participants, they also had to endure numerous bureaucratic aggravations and frustrations.

**Conclusion and Recommendations**

Participant accounts form the basis for several recommendations for IID health services. The first of such recommendations centres on multi-disciplinary and personalized care. Many researchers and health professionals have advocated the adoption of such an approach for the treatment of HD (e.g., Benjamin et al., 1994; Nance, 2007; Skirton, 2005). This approach is also
reflective in the services provided at the HDMC and HDRC. Participants were universal in their praise for such services, both in terms of the quality and depth of service, as well as how they were communicatively engaged as individuals by health professionals. Indeed, the latter point is exemplary of Frank’s (2004) discussion of dialogical and mutually respective relations between patient and client, which he sees as a cornerstone for effective healthcare.

Although this approach was a success within the confines of the HD clinic, it was not reflective of the participants' experiences with non-specialized medical services. In contrast, respondents noted that they were often ignored, mistreated or provided substandard medical service. Many health professionals were also not interested in consuming the consultation services provided by the HDMC, and showed little interest in learning about the disease. As such, interdisciplinary and individualized care appears dramatically less feasible within the context of broader healthcare. There seems little amelioration for health professionals who have no interest in learning about HD or in consulting experts. Continued education of health professionals who are not HD experts, as well as advocating for clients, will be an important step in meeting the standards for HD care outside of specialist clinics.

The second recommendation that can be drawn from participant accounts is the creation of additional forms of peer support. Such groups have long been identified as sites for emotive disclosure, information dissemination and health promotion (e.g., Docherty, 2004). The majority of respondents were avid consumers of such resources and accrued similar benefits. The HD peer supports also shared the model of other illness support groups, which orient around individuals who are physiologically ill and their partners (e.g., Coreil & Behal, 1999; Krizek, Roberts, Ragan & Ferrara, 1999). While this model may be appropriate for diseases such as cancers, HD has a number of specific ambiguities. In particular, participants who were asymptomatic, or were expressing low-level symptoms, felt as though their situation was unique in comparison to other HD sufferers. Although support group research has suggested that individuals at different stages
of the illness trajectory both have different needs and attend for different reasons (Oliffe, Halpin, Bottorff, Hislop, McKenzie, & Mroz, 2007), a stage-specific support group would be innovative. Huntington Disease researchers have already made several comparable innovations by highlighting the diagnostic, ethical and discrimination issues specific to low symptom individuals (e.g., Benjamin et al., 1994; Bombard et al., 2007; 2008; Cox, 1999). Therefore, the unique needs of individuals who are experiencing few, if any, symptoms has already been established. An important next step, as advocated by this study’s respondents, would be the creation of a peer support resource that incorporated the findings of these numerous researchers and was responsive to their particularities.

The third, and final, recommendation emphasizes the need for a disability system that is sensitive to the realities of HD. Although disability is a consequence of the degenerative nature of HD, few studies (cf. Roger, 2005) have engaged with this issue. However, the issues raised by respondents, specifically the loss of autonomy and the difficulties in accepting a disabled identity, are common experiences for individuals with other conditions (e.g., Ells, 2001; Hockenberry, 1995). Particularly resonant is Williams and Collins (2002) discussion of being transitioned from a person with an illness to a “significantly disabled chronic patient” (p.299) through the feedback of others, particularly health professionals. Unfortunately, there seems to be a dearth of resources available to help individuals respond to the rapid and frustrating recategorization from “abled” to “disabled”. Moreover, the actual disability forms seem to communicate an opposite message. Specifically, the forms are highly reflective of what Frank (1995) calls the “restitution narrative”. Such narratives focus on the transition from health, to illness, to recovery. As HD is an incurable, genetic condition, such messaging seems misplaced. Disability benefits appear to present somewhat of a paradox for people with HD. To access the funds individuals must have a lifelong condition and accept the term “disabled”; however, the
process of reimbursement itself emphasizes questions of recovery, an occurrence that is mutually exclusive with HD and gaining access to disability benefits in the first place.

The genetic and diagnostic features of HD provided additional challenges for disability regulation. Numerous reports have suggested that the cognitive and psychiatric effects of HD may appear years in advance of the motor effects (e.g., Duff et al., 2007; Marshall et al., 2007; Stout et al., 2007). However, it is the latter category that diagnoses are based on, and to access disability benefits, individuals must have a diagnosis. Accordingly, although individuals’ ability to work is compromised by HD symptoms, they are not provided disability benefits. Ironically, Canada’s disability policy states that the benefit is for those “whose disability prevents them from working at any job on a regular basis. The disability must be long lasting or likely to result in death” (Service Canada, 2009), criteria that HD is quite likely to fulfill. It appears that HD provides a number of difficult ambiguities for current disability services. Like the issues with peer support described above, these difficulties largely stem from the predictive genetic test, as well as the recent research on cognitive-psychiatric symptom onset. Similarly, another recommendation from this study’s participants would be the adjustment of disability forms and access to reflect both the extensive research findings and the realities of HD.

In summary, all of these recommendations highlight a tension between the nuances of HD and the generalizations of healthcare. Individualized care has been a success in HD clinics, yet has difficulty translating outside of their confines. Research has identified the unique needs and experiences of asymptomatic individuals, yet group support has not been adjusted for their needs. Likewise, disability regulations seem poorly adapted to both the genetic nature of HD, as well as the ambiguities of symptom onset. Individuals with HD consequently stand across a divide between their particular needs and standardized healthcare.
CHAPTER 4: THE ACCOUNTS OF INFORMAL CAREGIVERS

Introduction

Thus far, the experience of individuals with HD has been highlighted. At this juncture, the focus will switch to the experiences of informal caregivers. These individuals are the husbands, wives, siblings and parents of individuals with HD who have transitioned into a supportive role after their loved one’s diagnosis. Indeed, due to the hereditary nature of the disease, these individuals are often caregivers to several affected or at-risk family members. For instance, one participant was simultaneously providing support for her husband, son and granddaughter, all of whom had symptom onset. Given such broad familial consequences of diagnosis, HD has often been labelled a “family disease” (e.g., Brouwer-DudokdeWit et al., 2004; Sobel & Cowan, 2001). Previous research has also suggested that informal caregivers play an important role in HD care, arguably providing more support than formal healthcare services (Lowit & van Teijlingen, 2005). Accordingly, although HD physiologically affects specific individuals, there are numerous additional ramifications for family members and loved ones. Caregivers therefore provide another important avenue of investigation.

The experiences of caregivers have been central to numerous analyses of chronic illness. Their experiences are perhaps most ubiquitously associated with the phenomenon of caregiver burden, identified as feeling overwhelmed or fatigued by one’s caregiver role (e.g., Carretero, Garces, Rodenas, & Sanjose, 2009; Okamoto & Harasawa, 2009). Furthermore, caregiver burden is thought to be particularly salient for those tending to individuals struggling with dementia and cognitive decline, a common symptom of HD (e.g., Dunkin & Anderson-Hanley, 1998).

In addition to studies on dementia and chronic illness, caregivers’ experiences have also been central to analyses of fatal diseases. Indeed, a substantial amount of death and dying research focuses on the perspectives of loved ones, family members and informal caregivers.
(e.g., DesRosier, Catanzaro, & Piller, 1992; O'Brien, 1993; Scholteop Reimer, deHaan, Rijinders, Limburg, & vanden Bos, 1998). Of particular relevance to the study of how caregivers are situated vis-à-vis degenerative diseases is the notion of “social death”. This concept has been used to describe the process by which an individual is socially perceived as dead before they are biologically dead (Hertz, 1960; Kalish, 1966; Kastenbaum, 1969; Seale, 2000; Sudnow, 1967).

Specifically, Sweeting and Gilhooly (1992; 1997) have used the term to conceptualize how partners deal with fatal diseases with cognitive effects. In such situations, the authors noted that a substantial number of caregivers acted as though their still alive loved one was already deceased.

There have been a number of productive investigations into caregiver experiences, which discuss several potential difficulties that may be encountered by those engaged in HD care. These implications are perhaps compounded by the critical role HD caregivers play in the lives of their loved ones (Brouwer-DudokdeWit et al., 2004; Lowit & van Teijlingen, 2005; Sobel & Cowan, 2001). As such, after reviewing relevant literature, this chapter will analyze the caregiver experience from several vantage points. First, to provide additional nuances on HD, the accounts of caregivers will be presented on the content of the three previous chapters: death, psychiatry, and health services. The analysis will then switch focus to caregivers’ perspectives on their own role, centering on their feelings of isolation. Lastly, the analysis will conclude with caregivers’ thoughts on their absence of resources, and two recommendations to address this absence.

**Huntington Disease Caregivers in Health Research**

Previous research has explored caregiver experiences from a number of vantage points. As with HD research in general, one of the primary foci has been genetic testing. For instance, some caregivers, such as siblings, were formally at-risk individuals themselves, and research has suggested they may experience survivor’s guilt in response to their own negative test (Meiser & Dunn, 2000; Tibben, 2005). More commonly, caregivers are spouses or partners, with genetic
testing similarly having consequences for their lives. For example, Cox and McKellin (1999) noted that partners of individuals involved in predictive testing often carried the burden of communicating the results to other family members, particularly children. Similar findings regarding caregivers’ central role in communicating about genetic risk have also been reported by Forrest and colleagues (2003; 2005). Caregivers also appear particularly concerned about the ramifications of predictive testing for their offspring (Lowit & van Teijlingen, 2005; Williams et al., 2009). Accordingly, research has indicated that genetic testing affects reproductive decisions, with 100% of caregivers reporting they would have embryonic testing for HD (Decruyenaere, 2004). Positive test results may also decrease marital satisfaction (2007).

Researchers have also documented a number of the unique aspects of HD caregiving. For instance, Lowit and van Teijlingen (2005) suggest that the disease’s long duration and slow progression provide caregivers with time to develop adaptive strategies. However, other researchers have countered this claim by noting that the disease’s duration contributes additional burden as it creates a cycle of constantly reoccurring crises (Aubeeluck & Moskowitz, 2008; Skirting & Glendinning, 1997). Furthermore, research indicates that many caregivers need to reduce their working hours to handle their caregiving duties (McCabe, Roberts & Firth, 2008). With the HD’s long duration, this decrease in employment has implications for financial stability and retirement plans.

Studies have also documented the psychological difficulties encountered by HD caregivers. For instance, Williams and colleagues (2009) described caregivers’ experiences of “emotional disintegration” in response to their loved one’s diagnosis. Others suggest that caregivers experience levels of distress comparable to those who have personally received a confirmatory genetic test for HD (Decruyenaere, Evers-Kiebooms, Boogaerts, Demyttenaere, Dome & Fryns, 2005).
Many caregivers are also romantic partners of people with HD. As with genetic testing, symptom onset creates a number of unique issues for dyads. For instance, Kaptein and colleagues (2007) documented a disparity between partners’ and affected individuals’ reports of symptom debilitation. Specifically, partners perceived their loved one to be suffering from worse symptoms than the symptomatic individual self-reported. The authors also observed that quality of life measures were higher for both parties when symptom evaluations were similar.

The terminal aspect of HD also has ramifications for intimate partners. For instance, Dawson (2004) observed substantial distress in caregivers whose loved ones were receiving palliative care. She also noted that HD caregivers seemed particularly anxious about the future and appeared to have little information on the dying process. Additionally, similar to studies on the phenomenon of “social death” (e.g., Seale, 2000; Sudnow, 1967; Sweeting & Gilhooly), research indicates that HD caregivers anticipate the death of their loved one, and act as though the affected individual were no longer their intimate partner (Williams et al., 2009). An earlier study also highlighted that while caregivers witness exceptional changes and debilitation in their partners, their feelings of loss and grief are not socially validated, given that their partners are still biologically alive (Decruyenaere et al., 2005).

As such, regardless of whether they are romantic partners or not, caregivers encounter a number of difficulties. To cope with these stressors, caregivers adopt several strategies. For instance, many caregivers engage in emotional communication with a trusted friend or family member to both disclose their feelings and reduce distress (Roscoe, Corsentino, Watkins, McCall & Sanchez-Ramos, 2009). However, research suggests that HD caregivers primarily employ passive responses, such as avoidance and delaying medical appointments, particularly those that may confirm symptom onset (Decruyenaere, et al. 2005; Lowit & van Teijlingen, 2005). A study by Lowit and van Teijlingen (2005) also observed that caregivers both ignore and deny their own emotions in relation to HD. Unfortunately, these passive coping strategies are themselves
associated with increased distress, and decrease the likelihood that caregivers will themselves receive support opportunities (Decruyenaere et al., 2005; Lowit & van Teijlingen, 2005).

In response to the numerous difficulties faced by caregivers, as well as their employment of predominately detrimental coping strategies, researchers have made several health services recommendations. Healthcare professionals have been advised to be particularly attentive to HD caregivers, as they reported high needs compared to caregivers of similar diseases (e.g., Parkinson's Disease, Multiple Sclerosis) and also reported difficulties accessing health information and resources (Dawson, 2004; Kristianson, Aoun & Oldham, 2006). Health professionals have also been informed that caregivers are unlikely to attend HD support groups, as they experience difficulties viewing individuals with severe symptoms (Lowit & van Teijlingen, 2005). Nurses and clinicians have also been advised to regularly monitor the mental health of HD caregivers, as they experience episodes of severe distress (Williams et al., 2009).

In many ways, the experiences of HD caregivers are well documented. These individuals provide a substantial amount of support, experience great distress and have comparatively poor resources. The subsequent analysis follows these lines of investigation, while also providing commentary on unaddressed areas, such as suicide and challenges with healthcare bureaucracies.

**Caregiver Perspectives on Death and Suicide**

Caregivers interviewed for this study engaged with issues of death and dying on several levels. Although the majority of participants stated that they did not think about death, they were still concerned about the suffering and quality of care their loved ones would experience. Suicide was a salient issue for a few participants, who understood their loved one’s perspective, but encouraged them not to end their lives.

All the caregivers were quite open about talking about death and dying. However, the majority stated that they were not actively concerned with the topic and preferred to keep their
focus in the present. Julie (55) summarized this sentiment when she stated that she would consider the issue “when the time comes”. Likewise, Patrick (37) stated that he was just planning to “go with the flow”, as he and his partner had “five, ten, fifteen good years” before death was an issue. Importantly, this postponement of death did not equate to a denial of the issue or a complete avoidance of the subject. Instead, participants acknowledged that they were not thinking about the topic and, while it perhaps deserved more attention, it was difficult for them to engage with it and maintain a positive attitude. As Sabrina (53) stated,

Obviously he’s not lucky. He’s got a life sentence, I guess, and what we have to do is realize that we understand and know about this disease, so you have to live your life now and in the present moment because you know that something will change in the future. But, you know, we’re not quite sure exactly what that’s going to be but we need to live our lives, we only have this one time to do it.

Sabrina, like Patrick and Julie, emphasizes that she is focused on the present, and she and her partner are living in the moment. In contrast, she also acknowledges the ramifications of HD, by stating that her husband has a “life sentence” and noting that there will be challenges in the future.

These challenges were highlighted by numerous caregivers, who were very concerned about the suffering their loved one might later experience. Beth (60) discussed these issues by way of comparing HD to Amyotrophic Lateral Sclerosis (ALS):

I wish with Huntington’s they’d die really fast, because at some point it just goes on and on and on and it’s terrible, you know? So I think a disease like ALS is a much more kinder disease. I mean, that’s a terrible thing to say about such an awful disease, but it’s kinder in that the person doesn’t have to suffer for an undue length of time.

In addition to the difficulties involved in watching her partner suffering through a decade of slow degeneration, Beth added that having her husband live in a nursing home was a particularly difficult prospect. She was primarily worried about the quality of care that he would receive, as their ability to afford an exceptional nursing home was diminished by the economic costs of his early retirement. Other caregivers expressed a similar apprehension about the affordability of
palliative care, often adding in their difficulties with the technological prolongation of life. Rose (54) demonstrated her perspective on the latter point while discussing the condition of two of her husband’s HD affected relatives. She said,

[Husband’s] uncle, he was in fetal position for many years, yet they still fed him fluids, you know. My question is, is that quality of life? He has an aunt in [City], she probably weighs eighty pounds and she looks literally like a skeleton and it blows me away that she’s still moving around in their life, but she’s very demented and is just a hopeless little piece of human being sitting on the ground just fussing around with a piece of paper or her purse. It’s very degrading.

Rose drives home the point that the prolongation of life is sometimes unnecessary and inhumane. She contrasted this account to her experience in her country of origin, where people were much more “open minded” about euthanasia. Like Rose, it was this type of prolonged institutionalized suffering that most caregivers feared would be the fate of their loved one. Accordingly, although many caregivers stated that they were “focusing on the present”, they still expressed numerous apprehensions about end of life care. Both the financial costs and the misery their loved one may experience in a care home were particularly distressing.

Apprehension about end of life scenarios also contextualized the topic of suicide, which was primarily discussed as a way to avoid such prolonged suffering. In this sense, caregivers perceived suicide as somewhat pragmatic and as a form of euthanasia. While they understood this perspective, they also played an important role in convincing their loved ones to consider other options. Edna (46) communicated both of these issues in her account of her partner’s recent suicide attempt:

In August of last year, [Husband] attempted suicide by taking all of the medication, which was not something I ever dreamed that he would do. I guess he just got into a pretty depressed state and was really missing his sister [who died from HD] and I guess he did not know how to deal with those feelings. And the fact that he was symptomatic now and didn’t want to end up in long-term care. And it was to do with, he’s afraid of the dying process. I told him, “I can help you with a lot of things, I’m pretty smart and pretty logical, but this I can’t”. So, I know that [managed care] is coming and this has been so hard so far, and I know that it’s just going to get even harder.
Edna believes that her partner’s suicide attempt was a way to address his fears about dying and palliative care. She also added that his suicide attempts also brought her to her limits as a caregiver. Although she was his primary support for many other issues, including dealing with Obsessive-Compulsive Disorder and childhood abuse, his suicidal actions required her to access additional help. Accordingly, she arranged for them to attend therapeutic counseling together to help deal with some of the issues leading to his suicide attempt. As such, although she understood his motivations for attempting suicide and refrained from normative judgments, she managed to get him engaged in health services to address his suicidal ideations.

Natasha (37) shared a similar story. When she first met her partner, he was openly suicidal; this was connected both to his father’s HD-related suicide and his own concerns about death from HD. Although she personally does not believe in euthanasia, Natasha was quite supportive of his right to take his life. She said,

> At first when we discussed it I said well, “If you’re going to do it then, you know, you should have somebody there. I’ll be your friend and if that’s your decision”. At first, a few years back, I came to that acceptance, “if you want to commit suicide, that’s your business”.

However, after they decided to get married, Natasha informed her partner that she did not “want to see him go down that road [suicide]”. Although she was still adamant that it was his right to chose, she repeatedly expressed her preference that he not take his life. She further noted that her partner currently has a renewed vigor for life, and no longer appears to have suicidal tendencies. Indeed, she summarized her role in her partner’s life by stating that, had they not met, “he would be dead”. Both Edna and Natasha refrained from judging individuals who are suicidal, instead communicating an understanding of their perspective. However, it is also important to note that they both played critical roles in ameliorating their partner’s suicidal thoughts and actions.

Caregivers held contrasting perspectives on both death and suicide. While they stated that they were avoiding issues of death, they also communicated substantial concern about the dying
process. Likewise, while caregivers understood their partner’s motives for suicide, they also played a critical role in reducing their partner’s desire to end their lives.

**Caregiver Perspective on Misdiagnosis and Psychiatric Care**

The symptom progression of HD presented caregivers with a number of difficulties. Like both individuals with HD and psychiatrists, caregivers often mistook the symptoms of HD as mental illness. Additionally, the psychiatric aspects of HD lead to a number of trying encounters with the mental health system.

Caregivers experienced the effects of misdiagnosis somewhat tangentially, as few of their loved ones were directly misdiagnosed. However, several caregivers noted that individuals in their family, typically their partner’s siblings or parents, had expressed psychiatric symptoms. As many families were not aware of their history of HD, these individuals were often perceived as mentally ill. For instance, Beth (60) noted that clinicians thought her husband’s brother “had some kind of mental relapse, some kind of falling apart. And it wasn’t diagnosed for some time because there was no family history.” Sabrina (53) recounted a similar story about her husband’s brother:

We sort of thought he’d actually experienced a bit of a mental breakdown. And that’s what we took it as because he was just a little bit, you know, kind of bizarre in some of his behaviour. You could call it a bit of a break down because we didn’t think he had ever had those symptoms prior to that time, and we didn’t know what they were. So he just kind of became a different person. And then he started to lose interest in looking after himself he ended up becoming homeless and lived in his car and we had to intervene and have him find shelter. But again, we kind of chalked it up to, “wow, I guess he just has really lost it.” But there always was this question as to whether their dad had a mental illness. So that’s why I guess [husband] was diagnosed first.

Sabrina’s family and health professionals suspected her brother-in-law was manifesting a hereditary psychiatric disorder, as his father had expressed similar mysterious symptoms. Accordingly, her husband was the first to be diagnosed with HD, although he was the third
person in his immediate family to present symptoms. Like Beth’s brother-in-law, Sabrina’s in-
laws did not receive adequate treatment for their disease. Additionally, both of their partners, for
better or for worse, were not provided with advanced warning that they were at-risk for HD.

Other caregivers felt the effects of misdiagnosis more directly. No respondent elucidated
these problems better than Karen (59), whose son-in-law developed HD and had a psychotic
break. As a result, Karen was caring for the child her son-in-law had with her daughter, who was
unwilling to engage in childcare. At the age of two, Karen started noticing her granddaughter’s
difficulties in meeting developmental milestones. She began to suspect that her granddaughter
was also affected by HD, and she recounted the ensuing complexities of the diagnostic process.

As Karen stated,

I knew. She had behaviour difficulties, severe learning difficulties, inarticulate speech,
awkward gait when she was walking. I knew that’s what it was. She wasn’t diagnosed
until I guess she was 12 or 13. They thought she had ADHD, Tourette’s and
Oppositional-Defiance Disorder, but now we know it was all HD… If a parent is seeing a
problem with a child, and Huntington is a possibility, then I think it should be the
parent’s right to get them tested, because then you know what you are dealing with. I
mean, the scattered diagnoses that we had for her before that were just that, scattered
diagnoses. And the one thing that covered all of this was Huntington’s. And if we’d
known that in the first place. I mean, we had this poor child in remedial math classes, in
Kumon [learning centre], she was going to learning disabilities centres. If we had known
it was Huntington’s we wouldn’t have pushed all that stuff on her and made her unhappy
by forcing her to do this ridiculous homework.

Karen struggled to get her granddaughter evaluated for HD, even though the disease seemed a
strong candidate. She was told that, unless her granddaughter had a major medical problem, such
as a seizure, she was not eligible for a genetic test or diagnosis due to her age. Karen
subsequently followed the advice of health professionals, who suggested her granddaughter’s
condition was psychiatric and could be addressed with behavioural modification and learning
resources. In retrospect, Karen feels as though due diligence was not done in regards to her
granddaughter’s condition, and that the treatments she received were both unnecessary and
ultimately detrimental to her quality of life.
The accounts of Karen and Sabrina also serve to highlight the problematic nature of psychiatric treatments. Indeed, while other caregivers’ loved ones were accurately diagnosed with HD, they still had reservations about the subsequent psychiatric care. In particular, several participants encountered clinicians who were insensitive to the needs of their loved one. Edna (46) recalled a particularly combative psychiatrist who attended to her husband after he checked himself into a hospital, fearing that he was having a mental breakdown:

We first went to emergency and the psychiatrist came to talk with him and then started to talk to him directly about Huntington’s. And [Husband] got really agitated because he refused that this had anything to do with Huntington’s. So I hauled the psychiatrist out of the room and kind of gave him shit and said this is not the way to approach [Husband] about this. You are just going to agitate him further.

Edna suggested that the psychiatrist did not have an adequate understanding of HD and his decision to confront her husband about it during a psychiatric episode was ill advised. This decision is particularly ironic, as the psychiatrist does not realize that his approach is a source of psychological distress for his patient. After receiving feedback from Edna, the psychiatrist reframed his approach, and her husband was put on new medication and released from hospital shortly thereafter. While Edna’s account underscores problems with psychiatrists, it also emphasizes the important role caregivers can play in the health of their partners.

Jane also had numerous struggles with the psychiatric care available for HD. However, unlike other participants, Jane felt as though psychiatric professionals were exceptional in their ability to handle HD related issues. Instead, her frustrations focused on health bureaucracies and the absence of resources for individuals with psychiatric needs. Jane’s encounter with psychiatry started when her HD affected son had a psychotic break and began to believe that his head was being “fried with radiation”. He made the decision to check himself into hospital, much like Edna’s husband. This decision began an epic ordeal of dealing with the lack of psychiatric services available on Vancouver Island. She said,
He ended up getting admitted to the psychiatric intensive care unit at [hospital]. And he ended up spending 25 months in there. Which I was told by [politician] that he holds the record for length of time in there. I just proceeded to tell him that that wasn’t a record that anybody should be proud of having, or there shouldn’t even be that long of a record anywhere, for any medical problem. Anyways he eventually got on the waiting list for the only mental health tertiary care facility on the island, which is where he is now. So that’s the only place on the island. The government promised another one to be built in Nanaimo five years ago and they have yet to break ground on it. And that’s for the whole island. I mean it’s ridiculous since they closed Thorn Hill there’s no place for mental health patients. I have no complaints about the staff there, it’s the medical system.

Jane’s son spent 25 months on a ward intended for patients needing short-term, intensive psychiatric care. The unit did not have the resources to handle his needs. He was also able to escape from the premises several times due to poor security, at which time he would walk for several hours back to Jane’s house, where she was required to call the police to escort him back to hospital. These encounters with the police were particularly difficult for Jane. She further suggested that the health professionals also had deep concerns about the care they were able to deliver, and were trying desperately to get her son into the tertiary care centre. However, she reported that their ability to complain about service was limited by a government gag order. This order did not stop one doctor from resigning over these poor conditions, nor did it stop Jane from publicly speaking about the absence of mental health resources for people with HD.

Caregivers’ encounters with psychiatry highlight the challenges HD presents to family members, clinicians and the individuals with the disease. Caregivers noted that, even with an accurate diagnosis, psychiatric care was often not up to standards. Caregivers also reported systemic and bureaucratic problems influencing how psychiatric care can be delivered, as regulations affect both when a diagnosis can be made and what services are available.

**Caregiver Perspectives on Huntington Disease Health Services**

With caregivers’ reservations about palliative and psychiatric care reviewed, it is now appropriate to turn to their perceptions of HD healthcare. Caregivers had several concerns about
the services available to their loved ones. In particular, respondents commented on the venue for group support, doctors' communication skills, and difficulties in accessing disability payments.

Caregivers were universal in their praise for the staff at the HDMC and the HDRC. They appreciated being involved in their loved one's appointments and valued the dedication of the employees. The HD support group, operated by the HDRC and HSC received several positive evaluations, and while many caregivers did not attend the group themselves⁷, they made an effort to transport their loved ones to the meetings. However, while caregivers thought the group provided an emotional outlet and informational resource for people with HD, it was also the only HD specific service that they critiqued. These critiques centered on the venue hosting one of the support groups. The meetings took place in a nursing home, with the meeting room itself being well within the complex. Accordingly, attendees were required to walk through numerous areas of the nursing home to reach the group. While traveling through the home, participants noted that they frequently passed by, or interacted with, severely ill individuals. Rose (54) shared her perspective on this:

It's a care home. So you walk down this big long hallway, the place smells like urine. I mean there are people there that are elderly and ill and demented and everything and you see all these people being helped in the bathrooms and you know, shuffling around or half naked. A little old lady comes to us and asks if we can undress her and I think it's a very depressing sight.

In addition to engaging with symptomatic individuals at the meeting itself, Rose and her partner confront seriously ill individuals before the meeting even begins. This act was particularly disturbing for Rose and her partner, as one of his greatest fears was residing in managed care. Accordingly, although she found peer support beneficial, reaching the actual venue was quite traumatic. Rose mentioned that she was trying to have future meetings held in another location due to these difficulties.

⁷ This issue is discussed in the last section of this chapter.
Such comments about support group locations were the entirety of the complaints about HD specific services. However, like the individuals with HD themselves, caregivers had numerous reservations about external medical professionals, particularly in regards to their communication skills. For many individuals, these difficulties started as they were exploring the appearance of mysterious symptoms in their loved ones, as many were unaware that HD ran in their family. Numerous caregivers commented on the frustration of dealing with medical professionals who did not appear to acknowledge their loved one’s symptoms. For instance, Vicky (63) noted that, as her partner began to experience chorea, “he couldn’t sleep, he tossed and turned all night long”. When their GP was informed about these uncharacteristic movements he “just basically said ‘oh don’t you hate that when that happens?’ and gave him some sleeping pills.” Julie (65) and her partner had similar problems getting his symptoms acknowledged. She said,

For probably three years I kept saying, “there’s something wrong,” and the doctors didn’t really look at it. And, after he had the stroke of course he saw [doctor] and when we went to see him for that we just discovered that [Husband] had Huntington’s. When we walked in he said, “there’s something wrong.” And I said, “Yes, I know there’s something wrong, but nobody seems to be able to tell me what’s wrong.”

Both Julie and Vicky suspected that their partners were ill, but could not get medical professionals to take their observations and concerns seriously. As a result, their partner’s HD was not discovered until they developed additional symptoms.

Caregivers also reported similar communication difficulties post-diagnosis. These challenging interactions were primarily with GPs and specialists, such as surgeons, dentists, and anesthesiologists. A primary concern was that, although their GPs were responsible for medication management, caregivers often perceived them as having poor knowledge of HD. When trying to provide informational resources to their GPs, caregivers often encountered difficulty. As Sabrina (53) noted,
I find my role or [Husband’s] role is almost to educate the doctors. I’m sure there will be doctors who, in their lifetime of practicing, will never even have a Huntington’s patient. [Husband] and I saw a doctor who said “why did you bother coming and telling me this” and we said, “well maybe you didn’t understand about Huntington’s” and he was like, “oh no, yeah I knew a friend of a friend or someone who had Huntington’s” so he said, “I know about Huntington’s.” I sort of thought well, that may be true, but he had no empathy for what [husband] was trying to tell him, so that was frustrating.

After their partners were diagnosed, Sabrina and other caregivers thought it was important to provide GPs with a list of approved medication, as well as a book for physicians printed by the HSC. These actions were partially informed by the numerous difficulties both caregivers and people with HD encountered as a result of uninformed health professionals. Although many participants had doctors that expressed an avid interest in learning about HD and engaging the HDMC, just as many recounted stories similar to Sabrina’s. In the latter cases, caregivers suggested that the doctor was offended or slighted that their patient would offer them guidance.

In some instances, uninformed healthcare professionals and bureaucratic policies combined to create extremely challenging circumstances for people with HD and their caregivers. Edna’s account of her and her partner’s interaction with the Insurance Corporation of British Columbia (ICBC) and a cohort of doctors following a motor vehicle accident best exemplified these difficulties:

He was kind of slow and dizzy and had difficult buttoning up his clothes and the head injury really quite affected him for a while there. So the neurologist and other medical professionals couldn’t conclusively say that it was or wasn’t Huntington’s, even though his symptoms improved from the car accident. This was one thing I had to keep harping about. Like, HD symptoms don’t get better. And they also don’t all of a sudden happen overnight. He had severe head pains, which is not a symptom of Huntington’s, and the doctor didn’t even write that in the report. He had some bleeding out of his ears that the doctor said was a sinus infection, which was, coincidently [sarcastic], at the same time as the accident. His GP was an idiot. So, the symptoms from the car accident took quite a period of time to subside. And they just put it down to his Huntington’s, even though he didn’t have any Huntington’s symptoms previous to that. So it was a real fight that, of course, we didn’t win because you can’t win against ICBC. And they dragged it out so long that [Husband] spent all his savings and was basically forced to settle.
Although Edna’s partner was experiencing a number of symptoms, the car accident, inadequate evaluation, as well as the financial and legal pressures of ICBC, combined to define his car accident symptoms as HD symptoms. As HD is a “pre-existing condition”, her partner received little reimbursement for his accident, and ICBC manage to portray him as remarkably unharmed after his head-on collision. Consequently, Edna and her partner find themselves disempowered by a bureaucracy and network of health professionals, thus excluding them from benefits to which they should be entitled.

Bureaucratic policies were similarly described as excluding individuals from disability benefits. Specifically, many people with HD did not receive their benefits because the caregiver was still employed. As Patrick (37) said,

There’s also no help from the government supporting [Wife]. I mean, I don’t expect anything but, [Wife] is not getting any disability or whatever. I think I’m making about $3000 a month and I think the line is a really lower for that. Because, I mean, I don’t expect anything for myself but it’s just barely working out during the months for us financially. But something like Christmas, I have to go back to my savings and I’d actually like to save my savings for retirement.

Patrick exemplifies the difficult position that many caregivers found themselves in, namely that their partner’s disability prevented them from working, meaning a single individual’s wages needed to support two people. While this situation was tenable in the short term, as Patrick suggests, it decreases the dyad’s ability to have any form of savings or retirement funds. While this is problematic for the caregiver’s post-work quality of life, it also has implications for what type of managed care they can afford when severe symptoms appear.

Although caregivers were largely enthusiastic about HDMC and HDRC services for HD, they provided feedback on several aspects of HD care. Primarily, their comments highlighted the frustration of not being engaged as a human being by health professionals and bureaucracies. They also reveal the restrictive, and somewhat illogical, policies of these bureaucracies and the problems of capital they created.
Caregiver Accounts of Isolation and Loneliness

Thus far, this chapter has intersected caregivers' accounts with the themes of previous chapters to provide their perspectives on several aspects of HD and HD care. However, the disease also had consequences and repercussions for the caregivers themselves. In particular, although every caregiver was extremely dedicated to their loved one, they experienced a profound sense of isolation. This isolation was both connected to how the disease affected the person in their care, as well as its implications for other family members.

The sense of loss and isolation felt by caregivers often began at the moment of diagnosis. Many participants recounted feeling devastated after hearing of their partner's symptom onset, or inheritance of the gene for HD. Additionally, caregivers often suspected their partners had HD well before the individual noticed symptoms in themselves; this was typically informed by a movement or expression similar to an affected family member. This advanced knowledge was difficult for caregivers, and several acknowledged that they, perhaps intentionally, dismissed their observations. However, Beth (60) noticed the symptoms in her husband almost a year before he was diagnosed and decided to keep this knowledge to herself:

So I asked my best friend, “if you didn’t know you had a terrible disease and you weren’t going to know for a little while, would you want to know?” She said, “no”. So, I said nothing to [Husband]. It’s really kind of a nightmare for me because, for a couple of years, I didn’t say anything to him and then finally I said, “I think you need to get tested,” because it was becoming very much more obvious and obviously not a secret from him anymore. So we did get him tested and I think that, for him, he kept holding out hope until that day that we were told. I know that when we were driving there he said, “maybe I don’t have it,” and I said, “[Husband], don’t expect that’s what they’re going to say, because I don’t think there’s a hope you don’t.” For me there was no doubt, there was no doubt from the first time I saw one of the facial expressions he made, I knew. It was exactly like his brother. So there was absolutely no turning back and so it was no surprise for me. And in a way it was probably better, although it was absolutely hell for me those years. I couldn’t sleep, I thought, “what’s going to happen?”
Aside from the aforementioned friend, Beth kept her suspicions a secret until her husband’s HD became obvious. Although Beth notes that this process helped her prepare for her husband’s diagnosis, she obviously suffered through an extreme amount of distress. In her attempt to provide her husband with one last year without HD, she took the full burden of knowledge upon herself and dealt with the subsequent difficulties largely on her own.

The diagnosis also isolated caregivers in a much more tangible sense. All of the caregivers in this study had no chance of inheriting the disease themselves. As such, they were one of the few people in their families “safe” from HD. This status was particularly problematic for participants who were mothers. For instance, Jane (64), whose husband, son and granddaughter were already symptomatic, discussed her apprehension about her other son’s upcoming genetic test. She said,

He’s just starting to meet up with UBC now to go in and be tested himself. I don’t know what I’ll do if he tests positive too, because that will mean both of my children [have HD] and all of my grandchildren are at-risk. Normally Huntington’s is classified as 50-50, you have the 50-50 chance of getting it. In my husband’s family there were four kids and two of them got it and two didn’t. So I just don’t know. Emotionally, it’s very hard.

Jane references both the outcomes in her husband’s family and as the HD probabilities to suggest an optimistic result for her other son. However, this optimism is countered by her son’s still quite realistic chance of receiving a gene-positive outcome. In the event of such an outcome, Jane mentioned that she has “no idea” how she will be able to support so many affected and at-risk family members on her own. Similarly, Sabrina (53) echoed Jane’s attitude when she discussed her own fear of her children’s genetic status:

I would like to know, but of course I would like them to come away knowing that they are clear of it. I have a great fear of that actually. That’s probably my worst fear in my whole life so, it’s kind of like getting on a roller coaster, you want to get on but once you’re there, you wish you could get off but you can’t. So, yes, I wish, we could sort of step on that [genetic testing], but I’m running as fast as I can the other way. Right now I have a horrible fear of it just an absolute terror, I guess. I don’t know how to say it. Just fear, you know. Concern for everyone and a fair amount of guilt. I can honestly say
because, in the whole family situation I know that I am the only one that absolutely will never have it.

Sabrina compares her children’s potential genetic testing to being on a rollercoaster to communicate the lack of control she feels in the situation. She wants to hear that her children are gene-negative, so that she can stop worrying, but does not want to hear a gene-positive result. Consequently, although Sabrina will never develop HD, this in itself is problematic as she is free of concern for herself, but stricken by concern for others. Like many of the caregivers, her genetic status separates her from her family, and is thus a source of guilt.

The manner in which the disease changed an individual’s personality was also a source of isolation for caregivers. Participants often felt helpless as attributes of the person they loved evaporated in front of them. For instance, Julie (65) mentioned how her husband now felt as though he were a “house guest” when visiting her from his care home. She, as well as Sabrina and Beth, used death metaphorically to communicate the changes brought on by HD. Julie stated, “It’s pretty much like a death sentence really, because you know that from now on nothing’s going to get better, it’s only going to get worse. It’s frustrating. I’ve accepted the he’s not the person that I married by any means.” These statements of loss were commonly connected to their partner’s emotional desensitization and lost communications skills. For instance, Beth (60) stated about her husband, “he’s kind of flat lined with his mood, um, that’s probably the drugs, and he used to be a person that laughed a lot, that doesn’t happen very much. And the hardest part is he doesn’t talk anymore.” Edna (46) echoed this observation: “I’m an emotional person and I need a lot of affection and support and connection. So not always having that’s been really hard.” Although Rose’s (54) partner started to express more emotions post-diagnosis, she experienced similar problems:

This transition took place where he became more emotional. He became more emotional, but in some ways, of course, I am losing my friend and my partner. Because his communication, his comprehension is different. Maybe he’ll understand but it may take him twelve or twenty-four hours sometimes before I get an answer back.
Rose may be able to connect with her partner in new ways, but paradoxically the disease has inhibited his ability to engage with her. However, it was Vicky (62) who most succinctly conveyed the loss experienced by caregivers after their loved ones changed. She said,

"Life is quite quiet when you live with somebody with Huntington's because they don't initiate a lot of conversation and they don't initiate a lot of activities. So life can be quite quiet. It's lonely, and it's long and it's painful and it's full of loss, so, I think that probably sums it up."

Although Vicky was retired and spent the majority of time at home with her partner, she still felt a deep sense of loneliness and isolation due to his silence and lack of spontaneity. Indeed, some of her happiest moments were the occasions when her partner would spontaneously make her dinner or clean their house, as she stated, "but then he'll come back when you least expect it and you think 'oh! You're still in there!'". The joy that Vicky receives from such simple gestures further explicates what she misses in her partner, as well as her feelings of loss.

In addition to these feelings of grief, loss and isolation caused by witnessing a partner's decline, HD also had implications for caregivers themselves. In many ways, losing a partner to HD was almost like having lost a potential, or anticipated, self. The plans of many caregivers, such as travel and retirement, revolved around doing such activities as a dyad, and with one member of the dyad seriously ill, these future plans were lost. As Beth (60) stated,

"Because you kind of give up on all the normal progression of things. You know, you retire, you travel, you do this, you do that and all of that. That door is closed. And that is a hard thing to face. I don't think about it a whole lot because it's upsetting to think about. It's upsetting to think that, chances are, when I get older I'm not going to be like my parents. They have each other in their eighties and nineties and I'm probably going to be on my own. Or maybe not, I mean, who knows, but I'm suspecting I probably will be. That's not a nice feeling. And it's not a nice feeling to think that your future is very different than you originally anticipated your future to be."

For many caregivers, HD threw their future into doubt. Indeed, the disease seemed to obscure a period of their lives that was previously mapped out. Caregivers did not know when and if they
would be widows, or for how long they would be caring for their loved one. These dynamics created a number of ambiguities, which were touched on by Julie (65):

   My life has changed a lot because I’m neither single nor married. I go out with girlfriends and couples and that, but I am a single person and that’s the big difference. I think most counselors, or whatever, say that [HD] is almost like death, you’ve lost that person and you get used to it. You know, my life, it has totally changed from what I thought it was going to be, as has [Husband’s]. I just am working my way through having just retired now. I am having to work my way through, what am I going to do with the rest of my life? How is my time going to be filled?

Julie’s account emphasizes the predicament that many caregivers found themselves in. Specifically, they felt as though they were on a precipice between two different lives, one of which centered around their ill partner and another that acknowledged that their partner was, in many ways, already gone.

   These accounts emphasize the loss and isolation experienced by caregivers. Although HD physiologically affected their partners, caregivers suffered a great deal of distress. Indeed, by being physiologically spared by the disease, caregivers often felt unique anxieties. As the disease progressively took their partners in a biological sense, they lost their partners, and an aspect of themselves, in a relational sense.

   Caregiver Resources and Recommendations

As has been shown, caregivers both play an important role in HD care and are profoundly affected by their loved one’s diagnosis. Although caregivers were grateful for the numerous resources provided to their loved one, they felt overburdened by their obligations. Accordingly, caregivers thought that there was a comparative absence of resources to support them in their new role.

   The increasing disability of their loved one had serious practical implications for caregivers. Most notably, they had to find a way to meet all the duties previously fulfilled by
their partner, while also providing care and support. Accordingly, as their loved one’s symptoms increased, so did caregivers’ workload. As Beth (60) noted,

It gets increasingly more difficult for me to do my job, run the house, the garden, everything. There’s so much [Husband] used to look after, which he kind of does a bit of, but you know, nothing like he used to. I can ask him to do things and for the most part he does, but he forgets and he has to be reminded and doesn’t do anything very quickly. I’ve certainly found this, is that I realized at this point in life that it’s only me, there’s absolutely nobody that’s going to hold me up but me.

Diagnosis created a litany of new roles and responsibilities. Being a caregiver certainly was a prominent new role, but, as Beth highlights, these individuals were also the primary wage earner, the cook, the cleaner and the organizer. This abundance of obligations had consequences for the individual’s stress and social life. As Beth notes, this situation was exceptionally isolating.

The loss of a partner created new burdens, and caregivers perceived there to be a dearth of resources to help them adapt to this situation. This lack of resources was particularly poignant compared to the number of HD services for their loved one. For instance, Beth noted that while she and her husband meet with mental health professionals, “they’ve never said to me, ‘do you need to see somebody? Do you need to talk to someone?’ No one’s ever said that or suggested that.” Vicky (62) expressed a similar sentiment when she recalled the reaction of a HDMC mental health professional to her marital problems: “[doctor] just got up and opened his office door and he said ‘nope, still doesn’t say marriage counselor’”. Both a doctor and a nurse similarly dismissed her when she requested they send a letter to her husband informing him that he needed to visit the hospital. Although her husband refused to go under her guidance alone, the hospital staff refused to offer her support. However, Patrick (37) best expressed the absence of caregiver resources and support:

[Wife] is waiting all day long for me to come home. I have to put my own needs to the side. It’s coming really to a point where I am like, “it’s too much for me.” I mainly communicate with UBC if they have problems with [wife], in that sense they are really supportive. But I mean, for me as a caregiver, personally, I have actually barely any resources. Barely a single one.
Patrick's account typifies the position of many caregivers. He acknowledges, and is grateful for the resources available for his wife; however, he has “barely any resources”, which is problematic in regards to his intense involvement, similar to other caregivers, in the care of his partner. That is, like the precarious financial situation of many caregivers, this situation may be tenable in the short-term, but does not appear to be so in the long-term.

While caregivers outlined their burdens and the absence of caregiver specific resources, they also made recommendations for health services. As can be gleaned from the comments above, better communication with health professionals is needed. However, caregivers were also particularly interested in psychosocial support. Indeed, many caregivers were receiving some form of therapeutic counseling to help them deal with their loved ones’ diagnoses. Accordingly, there were a number of explicit calls for psychosocial support for caregivers. For participants on Vancouver Island, this need was in the form of health professionals who could engage in crises management. Jane (64), for instance, noted the difficulty and guilt that resulted from contacting the police to help her deal with her son’s psychiatric episodes. Vicky (62) similarly stated the need for crisis management support, while also touching on the difficulty of including the police. She said,

>You know what would be really helpful is like a fulltime social worker that knew about Huntington’s. Because sometimes you can get into a crisis situation really quick. And I remember thinking that first summer we were here, who would I phone if I needed somebody? Who would I phone? Who would understand? Because you don’t want the police coming full bar into what they think is a domestic dispute. Because it’s deeper than that it. I mean it might be a domestic dispute, but the cause is different. We need a full-time social worker or a full-time psychologist.

Vicky addresses her concerns about entering a crisis situation while simultaneously pointing out her lack of support if such a situation should develop. In particular, her only recourse may be the police, which she fears may perceive the situation more as a criminal transgression than a medical issue. She therefore concludes, as did other participants from Vancouver Island, that an
HD specialist would be of particular benefit. Interestingly, the participants in the Lower Mainland, who do have access to such a social worker, did not mention this issue.

Instead of crisis support, the participants in the Lower Mainland were particularly interested in a caregiver support group. While such a group exists on Vancouver Island, there have only been occasional “caregivers only” support sessions in the Lower Mainland. A number of caregivers were interested in such support, while also expressing apprehensions about the current format. These caregivers reported that they would not consider attending the support group due to its time and location. As several of the caregivers worked full-time jobs, they could not make the Thursday afternoon meeting. Additionally, the site of the group was more than an hour’s travel for a number of caregivers, further reducing their ability to attend. Additionally, caregivers were especially apprehensive about attending a support group with highly symptomatic individuals. For instance, Julie stated that, “I find the group really depressing and I don’t want to know how bad it’s going to get. I can live with it today, but I don’t want to think that at some point he’s going to be in really bad shape” Natasha (37) expressed this anxiety in more detail:

I’ll be honest with you. I’m afraid I’ll find it very depressing myself. So probably that’s the underlining tone for me, I just don’t want to. I know it’s hard, I don’t want to see other people sick. I don’t want to get sad about it. Just seeing other people go through it, I don’t think I have that. I think it would be tough. I will keep happy and keep [Husband] very happy, because that keeps him youthful, it keeps him very optimistic.

Both Natasha and Beth perceive being “optimistic” and non-disease focused as central to maintain their caregiving role. Their apprehension around attending the support groups centres on their belief that viewing severely ill individuals will derail that optimism. As discussed above, caregivers who did attend the support group expressed similar concerns.

Importantly, caregivers such as Beth, Julie and Natasha did report that they would be interested in attending a caregiver-centric group. Participants who participated in one of the few
caregiver-focused support group meetings also emphasized the benefits. Patrick (37) expressed these benefits:

One day we actually had the opportunity to split the group into patients and caregivers. And that was actually quite nice because, there’s a lot of things inside of you working which are not coming out, or you don’t think about it. And I remember sitting down there and thinking, you know, in Germany we say “we have a dumpling in our throats”, and this means you are close to your tears. And I remember this other person, and she had been dealing with Huntington’s quite a bit longer than I have, and she all of a sudden burst out into tears. So everyone kind of had this thing inside themselves. In the beginning it was sort of a survivorship, or a sort of a strong energy, but then suddenly everyone felt so relieved, and people started laughing you know? It was actually quite a relief. So I think I’d like to find something like that.

For Patrick, the group offered him important emotional support and a cathartic outlet. He also emphasizes the difficulties of being an HD caregiver by comparing the position to being a survivor of the disease itself. Patrick added that he had placed a request at the HDRC for a regular caregiver support group. In the meantime, he was attending a general caregiver’s support group in another area of the city. However, his partner had a great deal of difficulty being separated from him during that time. As such, having coordinated support groups was by far the best option for Patrick and other caregivers.

These accounts underscore the numerous sacrifices that caregivers make for their loved ones. They also emphasize the important role they play in HD care. While caregivers were pleased to be included in their loved one’s care, they felt there was a lack of resources to address their own needs. In particular, caregivers desired more psychosocial support.

**Conclusion**

The accounts presented here provide further confirmation for many of the findings of previous caregiver research. Huntington Disease caregivers clearly play an important role in managing HD care (Lowit & van Teijlingen, 2005). Caregivers also experienced substantial caregiver burden, much of which was attached to their loved one’s cognitive decline (Carretero et al.,
Furthermore, caregivers reported a great deal of emotional distress after their loved one’s diagnosis (e.g., Aubeeluck & Moskowitz, 2008; Decruyenaere et al., 2005; Kaptein et al., 2007; Williams et al., 2009).

These findings also offer some adjustments to previous research. First, caregivers did predominantly cope with the diagnosis by employing avoidant strategies, such as avoiding both group support and the subject of death (Decruyenaere et al., 2005; Lowit & van Teijlingen, 2005). However, the use of these strategies needs to be contextualized both by the incredible emotional burdens involved with caregiving, and the relative absence of resources available to caregivers. Furthermore, caregivers made several recommendations for services that they would be interested in engaging with. Until such supports exist, it is perhaps unsurprising that caregivers avoid more intense engagements with HD.

The findings also provide context to the phenomenon of “social death” (e.g., Seale, 2000; Sudnow, 1967; Sweeting & Gilhooy, 1992). Many respondents used terms such as “like death” and “death sentence” to describe their partner’s condition. However, a critical difference between their stance and that employed by the participants in Sweeting and Gilhooy’s (1997) study is that the HD caregivers never treated their partner as though he or she were dead. Instead, the respondents used “death” metaphorically to communicate both their loss and their transition from “married” to a quasi-widowhood.

The accounts of caregivers are also similar to many of the accounts of people with HD presented earlier in this thesis. In particular, both groups understood why suicide was considered by some individuals and refrained from passing normative judgments. Conversations on suicide suggested how important caregivers could be in reducing suicide ideation, which is a well-demonstrated finding in terms of men’s relation to suicide (Gove, 1972). Both cohorts also had difficulty engaging in group support in its current format. Participants instead suggest that there
should be three distinct forms of group support: symptomatic, low and pre-symptomatic, and
caregiver.

Including caregivers’ perspectives also provides a number of critical observations not
present within the HD cohort. Specifically, caregivers connected several problems in HD care to
larger systemic issues. For instance, psychiatric misdiagnosis was linked to the regulations in
place for diagnosing minors. The role of the clinician in poor psychiatric care was backrounded
in comparison to the government’s inability to provide and organize psychiatric resources.
Lastly, the fears that both cohorts held in relation to end of life care were contextualized by the
progressive evaporation of resources occurring over numerous years of living off of a single
income while supporting a chronically ill partner. Without substantial capital, and without aide
from disability benefits, caregivers feared that their choice of home care would be constrained
more by economics considerations then quality of life. Accordingly, including the perspectives
of caregivers enables a better understanding of the disease itself.

In closing, HD is often termed a “family disease” (Brouwer-DudokdeWit et al., 2004;
Sobel & Cowan, 2001). The familial quality of the disease is readily demonstrated by caregivers’
importance in supporting individuals with HD. It is further demonstrated by their own feelings of
helplessness and isolation resulting from the disease. Indeed, in many respects caregivers
experience the disease as validly as the individuals who are physiologically stricken. Caregivers
are also financially drained, experience substantial distress, and arguably lose both themselves
and their partners to the disease. Caregivers appreciated being involved in the management of
their loved ones’ HD, but felt as though they were not supported and lacked the resources needed
to deal with their situation. The importance of these individuals in providing for people with HD,
in combination with the lack of resources to help them cope with their litany of new roles,
suggests a contradiction in care. Namely, while in terms of burden, HD may be a family disease,
it is not a family disease in terms of support.
DISCUSSION AND CONCLUSION

Summary of Findings

Previous HD research has focused primarily on issues involving genetic testing (e.g., Bombard et al., 2007; 2008; Cox, 1999; 2002; 2003; Cox & McKellin, 1999). This thesis sought to add to this body of knowledge by investigating experiences outside of HD testing. Findings emerged in four primary areas: attitudes towards death and suicide, experiences with psychiatry, interactions with health services and bureaucracies, and the needs and experiences of caregivers. These findings, reviewed briefly below, were described from the perspective of both caregivers and individuals with HD.

As discussed in Chapter 1, although cognizant that they had a fatal illness, people with HD refrained from fatalistic interpretations of their lives. They emphasized that they were not doomed to death from HD, stating that a number of other random circumstances could instead end their lives. However, similar to Simmel’s (1918/2007) essay on death, people with HD also felt as though they were more proximal to death than the average person. As such, their accounts emphasize how death may be salient long before the physiologically terminal stages in which it has typically been studied by social researchers (e.g., Exley, 2004; Walter, 1993).

Participants also shared their experiences and attitudes towards suicide, a topic that has received an abundance of attention in the HD literature (e.g., Farrar, 1986; Paulsen et al., 2001; 2005). A minority of participants stated that they had suicidal thoughts or actions; however, the majority of respondents conveyed empathy and understanding of those individuals who had taken their own lives, or were considering doing so.

Recent research has suggested that the cognitive and psychiatric features of HD may appear years before the telltale neurological symptoms (e.g., Marshall et al., 2007; Morris, 1991; Solomon et al., 2007). This symptom progression has consequences for how HD is diagnosed
and misdiagnosed. As reported in Chapter 2, numerous participants recalled a family member’s HD symptoms being misinterpreted as a mental illness, with one participant having experienced such a misdiagnosis herself. While highlighting the consequences of misdiagnosis (e.g., mistreatment), their accounts also emphasize the divide between neurology and psychiatry (e.g., Cunningham et al., 2006; Price et al., 2000). In contrast to recent research arguing the increasing neurologicalization of psychiatry (e.g., Rose, 2007; Whitaker, 2002), this thesis described the continuing practical disjuncture between neurology and psychiatry by utilizing Latour (2005) and Bakhtin (1986) to focus on the observational and discursive attributes of the diagnosis.

Participants’ assessments of health services were discussed in Chapter 3. Overwhelmingly, participants described the services provided to them at the HDMC and HDRC as exceptional. These positive reviews support current HD health services models emphasizing integrated teams of multidisciplinary professionals (e.g., Nance, 2007; Skirton, 2005). However, reports on HD services have not discussed clients’ interactions with outside health professionals. This thesis addressed this gap by describing the numerous difficulties participants had with various health services. First, individuals with few or no symptoms found it difficult to engage in current HD support groups, as they felt these groups focused on the experiences of symptomatic individuals. Second, participants reported a number of encounters with doctors who refused to learn about HD and ignored treatment recommendations from the HDMC. Third, participants found disability regulations to be poorly suited for people with HD, particularly in relation to the permanency of the disease, ambiguities of diagnosis, and the need to self-identify as disabled. These difficulties subsequently translated into three participant-based recommendations: 1) the need for stage-specific support groups, 2) the need for better communication and understanding from health professionals outside of the HDMC and HDRC and 3) the need for disability services that are more responsive to the realities of HD.
Informal caregivers also commented on these three areas, as discussed in Chapter 4. Caregivers expressed deep concerns over their loved one’s future suffering and diminished quality of life, while also contrastingly emphasizing the importance of remaining present-focused in relation to issues of death and dying. Like people with HD, caregivers communicated empathy and understanding towards individuals who engaged in suicidal thoughts or actions; however, as previously noted in suicide literature (e.g., Gove, 1972), caregivers also ameliorated such actions.

Caregivers experienced the misdiagnosis of HD by association. Like mental health professionals, caregivers recalled interpreting the early signs of HD as symptoms of mental pathology. Importantly, in contrast to the HD cohort, caregivers also described a number of additional systemic difficulties regarding the psychiatric treatment of HD. For instance, one participant noted that her loved one’s misdiagnosis was a result of age limitations regarding HD testing and diagnosis, whilst another detailed the absence of resources available to individuals with psychiatric needs.

Similar to people with HD, caregivers extolled the virtues of the HDMC and HDRC staff. Unfortunately, they also echoed the HD cohort’s concerns about other health services. In particular, they reiterated the difficulties of seeing visibly ill people at support groups, as well as health professionals’ lack of empathy and understanding. They also described their difficult encounters with bureaucracies and disability regulations. These encounters had negative effects on their long-term financial security, which was also impacted by their need to reduce working hours to accommodate their caregiving duties (McCabe, Roberts & Firth, 2008).

The experiences of caregivers themselves, also reviewed in Chapter 4, were the last area of investigation. As has been found elsewhere (e.g., Aubeeluck & Moskowitz, 2008; Decruyenaere et al., 2005), caregivers reported substantial emotional distress and isolation. On a genetic level, the disease isolated caregivers from their families, as they were not at risk for
developing HD. On a symptomatic level, HD isolated caregivers from their loved one, who they perceived as slowly disappearing as a result of the illness. Many caregivers compared HD to death in order to convey their sense of loss and isolation; however, unlike previous research on social death (e.g., Sudnow, 1967; Sweeting & Gilhooy, 1992), all of the caregivers were still deeply involved in, and concerned with, their loved one. In addition to losing their partner, many caregivers also expressed a loss of a future/anticipated self. That is, with their partner affected by a degenerative illness, many of their own plans and goals for their lives were disrupted. These feelings of isolation and loss translated into the caregivers’ critiques of current health services. In particular, they stated the need for a caregiver specific support group that was flexible to their working situation. Caregivers on Vancouver Island highlighted their need for a full-time HD social worker/psychologist to deal with crises.

**Analytical Summary**

In addition to the descriptive summary presented above, it is also important to provide an analytical summary of this thesis. Specifically, it is critical to comment on a theme that has consistently emerged when evaluating the previous chapters’ findings as a whole. This theme, as suggested at various times throughout this thesis, is the ambiguity that HD presents for a number of categories and concepts.

Numerous social researchers have commented on the productivity in investigating such category disruptions. Bourdieu (1992) went so far as to claim that category slippage was the “sociologist’s bread and butter”. A number of noteworthy empirical investigations have analyzed the boundaries around categories, such as Smith’s (1975; 1983) discussion of psychiatry’s rearrangement of patient narratives to fit diagnostic categories and Rose’s (1999) description of how borders and statistics were used to create populations, rather than describe them. Derrida (1998) is perhaps most ubiquitously associated with category disruptions, with his numerous
investigations into the ambiguous space between categories that are apparently exhaustive and mutually exclusive poles, such as “alive” and “dead”.

These theorists highlight the difficulties in conceptual categories, a problem which will now be detailed in reference to HD. Borrowing from Said’s notion contrapuntal reading, this summary will seek to describe the ambiguities within categories raised by these participants, rather than suggesting new categories or refining old ones. In his work *Culture and Imperialism*, Said (1993) discussed the concept of contrapuntal reading, wherein imperialist depictions are read in counterpoint to their unaccounted for opposites. Said borrowed the term from music, where it was used to describe two notes that appear as counterpoints to one another, and are therefore both emphasized. Returning to the musical origins of the term, it is with similar intentions that the contrasting perspectives appearing throughout this thesis are placed alongside, or in counterpoint, with one another. It is not the aim of this summary to argue that one of these perspectives is somehow more authentic or accurate then another. Rather, it is to merely highlight the various ambiguities and different reactions that HD elicits from both individuals and organizations, while providing a closing synthesis of this thesis’ findings.

These contrasting positions are perhaps most clearly seen in participants’ discussions of death. For instance, in Chapter 2, participants noted a tension between “knowing” and “not knowing”, as well as both a proximity and distance to death. Additionally, participants viewed their lives as open-ended and meaningful, but several also considered suicide. Phyllis demonstrated the latter tension, as she both anticipated the joy and love of a new child, whilst also considering taking her own life.

These tensions are further highlighted by relating the accounts in Chapter 2 to those of the caregivers in Chapter 4. The former chapter raised questions of temporality in relation to social science studies on death, with the caregivers’ statements of HD being “like death” or “like a death sentence”, further emphasizing these questions. That is, while individuals with HD stated
that the disease did not finalize their lives, many caregivers felt that the disease did impose some finality. In several cases, members of the same dyad, individuals who had spent the majority of their lives living together, expressed these contrasting perspectives. Placing these contrasting accounts alongside one another further questions the “when” of death and dying, as there is an apparent disjuncture between caregivers and individuals with HD in regards to when these issues become relevant.

Importantly, caregivers also used death metaphorically to communicate their own grief, loss and isolation. This usage was in turn connected to caregivers’ discussions of their own lost life, namely their anticipated future, to which their partner was essential. Indeed, it appears as though issues of death are arguably as immanent for caregivers as for people with HD, both in terms of their lost partner and their lost self. Placing these two perspectives in dialogue adds further ambiguity to discussions of death, particularly around who experiences death and dying.

Another resonant contrast relates to how the participants and caregivers categorized themselves. One area that these categories become particularly salient is in regards to participants’ statements on their preference for support group membership. For instance, although the majority of caregivers were members of a dyad, they preferred to attend a support group with other caregivers, suggesting that the caregiver’s experience of HD is distinct from that of the individual with the disease. On the surface, this division between caregivers and individuals with HD is quite logical; indeed it is also utilized within this thesis. However, it is important to note that many of the individuals with HD were also caregivers. For instance, Keith tended to his older brother and Kenneth cared for one of his daughters. Furthermore, while not a reality in this study, many HD caregivers are previously at-risk individuals themselves. For example, I informally met with one individual who was the only gene-negative member of her immediate family. While she was currently their caregiver, she too had previously lived with the prospect of having HD. By underscoring these overlapping roles, we can highlight the
ambiguous terrain between “caregiver” and “person with HD”. Separating individuals into these two groups does appear useful for both parties, as well as for the purposes of this research; however the grey areas between their experiences are perhaps reflective of the nature of a “family disease”.

In a similar fashion, the individuals who were presymptomatic and those with few symptoms perceived their experiences as distinct from those with more severe symptoms. These individuals emphasized the need for a support community of individuals who were currently having similar disease experiences. In particular, they argued that their needs and perspectives were different from those of individuals with noticeable chorea. However, individuals with chorea did not divide disease experience along symptom milestones. Prima facie, the distinction between presymptomatic and symptomatic is also quite intuitive. As mentioned in the introduction, individuals who are gene-positive do not have HD, as they can only be said to have the disease after a neurological diagnosis. However, as previously discussed, an abundance of evidence suggests that cognitive and psychiatric symptoms may appear before the telltale neurological markers (e.g., Duff et al., 2007; Marshall et al., 2007; Stout et al., 2007). Consequently, participants’ distinctions along the basis of symptom presence and progression may become increasingly ambiguous as biomedical research advances.

The effects of these recent findings on symptom progression (e.g., Duff et al., 2007; Marshall et al., 2007) also underscore ambiguities in relation to the diagnosis. Issues of diagnosis and symptom onset were covered in Chapter 2. Huntington Disease appears to fit into psychiatry’s diagnostic categories, when in fact it does not. Grace’s misdiagnosis exemplifies this disjuncture, as she was originally perceived to have had a psychiatric disorder. Importantly, Karen’s account of her granddaughter’s misdiagnosis reveals how psychiatry is itself constrained by the policies of other institutions. In this case, a psychiatric misdiagnosis was the only
diagnosis available, as healthcare policies prevented her granddaughter from receiving a diagnosis of HD.

Indeed, Karen’s case also demonstrates another problem of HD diagnosis in organizational settings. Benjamin and colleagues’ (1994) policy paper provides the guidelines for predictive testing. Due to the psychological difficulties that testing may cause, these guidelines recommend individuals reach the age of majority before they can be tested. While this policy is helpful and protective of the majority of individuals with HD and their families, Karen’s granddaughter’s JHD raises ambiguities within these policies. Specifically, Karen argues that the inability to get a genetic test, and therefore a neurological diagnosis, was more damaging to her granddaughter than the psychological consequences involved in testing. Critically, the difficulties that Karen detailed were related to the fact that severe symptoms had already started for her granddaughter, whereas genetic testing policies are predominately geared towards individuals expressing a more typical HD trajectory.

The ambiguities of HD symptom onset also created difficulties for disability providers. As was described in Chapter 3, Phyllis experienced cognitive and psychiatric symptom onset, but did not yet meet the criteria for a neurological diagnosis. Being unable to secure this diagnosis, she could not receive disability payments, yet her symptoms also prevented her from performing tasks necessary to maintain employment.

Bringing together the accounts of Grace, Karen and Phyllis illuminates the ambiguities and contradictions that HD creates for disciplines, healthcare policies and organizations. However, also resonant within these accounts are the ambiguities that HD holds for neurology. Namely, the initial symptoms of a disease that neurologists’ diagnose does not manifest under the rubric of neurological diagnostics. That is, a neurologist, particularly one that knows an individual’s genetic status, is placed in the difficult position of being aware that an individual is expressing symptoms of a disease, but is unable to diagnose the individual with the disease.
Thus, the individual is sick, but does not receive the authorization to be recognized as such. Perhaps stated more clearly in Kleinman’s (1978) terms, the individual may be experiencing the disease (illness) and others may see that they are ill (sickness), but they are not identified as medically sick (disease). Grace, Karen and Phyllis’ respective dilemmas provide exceptional demonstration of these contradictions and complexities, as they, or their loved one, could not get adequate resources without being medically defined as ill.

By attending to the numerous counterpoints within the accounts of people with HD and their caregivers, we can explore the ambiguities, such as those created around diagnosis, that HD presents both for those who study it and those who experience it. Returning to Said (1993), it was not the goal of this summary to resolve these tensions, but to both reveal and emphasize them.

**Study Limitations**

Before concluding, it is important to detail the limitations of this thesis. The first major limitation of is its generalizability. Numerous authors have discussed this issue in relation to qualitative research, with Becker’s (2001) commentary standing as an exemplar, noting that qualitative research aims for detail and depth of description. However, it is still important to note that the project contained only 20 individuals with HD and 10 caregivers, representing only 5% of the individuals on file at the HDRC (S. Tolley, personal communication, June 2nd, 2009). Consequently, the attitudes, perspectives and experiences of these individuals may not be illustrative of the larger HD community in British Columbia.

Another important limitation to this study is its regional specificity. There are nuances between the healthcare policies of each province. Furthermore, each province has different access to research professionals. British Columbia is unique on a number of levels. First, the Vancouver HDRC has the largest portfolio of individuals with HD. This high population of
individuals with HD is in turn related to the second point, that UBC retains a community of leading HD professionals. As such, the individuals in British Columbia are receiving advanced care, partaking in new studies and have access to a larger community of individuals with HD. In another province with less research money and fewer individuals with HD, the experiences of individuals with the disease may be markedly different.

In a similar fashion, features of Canada also contextualize these findings. As researchers have noted (e.g., Hayden, 1981; Wexler, 2008) the experiences of individuals with HD may vary quite dramatically from country to country, with Wexler’s (2008) discussion of the exceptionally respectful way in which individuals with HD were treated in Venezuela providing a particularly good example. Furthermore, the experience between countries is influenced by different healthcare regimes. It would be a mistake to suggest that the experiences of this study’s participants would be comparable to a country like the United States of America, where there are different costs and levels of access to healthcare. Additionally, with particular reference to Chapter 2, the experiences of these participants may not comparable to individuals in Europe, as European countries use a more integrated diagnostic tool (the ICD-10), as well as having socialized healthcare.

The study is also limited in terms of the type of data that it draws on. This study was based in qualitative methods and, as such, is focused around the accounts of individuals and their experiences. As a number of anthropologists and researchers who utilize narrative analysis have observed, individuals employ multiple stories, which change in reference to audience, time and context (e.g., Briggs & Mantini-Briggs, 2003; Frank, 1995). Under different contexts, environments and researcher-participant interactions, these stories may be distinctly different. Accordingly, with a change in researcher or a greater distance between events, the perspectives of this study’s participants may change. This is not to say that one story is more authentic than another, as each account is authentic in its own right. Rather, it is important to highlight that the
interpretations provided by these study participants are just one of many of their valid
interpretations of events.

**Future Research**

As is evident by counterpoints and ambiguities that were reviewed above, there are a number of
areas for future investigation. Foremost, more research is needed to further address a number of
the new areas explored by this thesis. For instance, the issue of psychiatric misdiagnosis
commands greater investigation. As researchers have pointed out, 25% of individuals are not
aware that HD runs in their families (Almqvist, Elterman, MacLoed & Hayden, 2001), and, as
suggested in Chapter 2, may be particularly likely to be affected by misdiagnosis. Future
research should address both the effects and prevalence of such psychiatric misdiagnoses.

Given the diversity of experiences vis-a-vis HD, future research should attempt to
involve as many shareholders as possible. Specifically, as discovered in my experience at an HD
camp (see Appendix 1), the perspectives of HD care professionals were markedly different that
those of individuals with HD. For instance, in contrast to this study’s cohorts’ positive evaluation
of HD-centric resources, care professionals perceived that they were unable to meet the majority
of their clients needs. Future research should explore HD care professionals alongside
individuals with HD and their families.

Comparative studies are also another area of productive exploration. For example, this
study originally sought to compare the experiences of individuals with HD to people with
Amyotrophic Lateral Sclerosis (see Appendix 1). Such a project would be able to speak to the
nuances within degenerative diseases, such as the different symptoms and life expectancy.
Furthermore, comparative studies that focused on neurological diseases that activate different
disciplinary boundaries would also be important. As discussed in Chapter 2, Alzheimer’s Disease
is within the DSM-IV, while HD is not. Accordingly, a possible area of investigation could focus
on how these two diseases are conceptualized in relation to the spaces between neurology and psychiatry.

Another area of potential investigation would be the experience of HD under different healthcare regimes. For instance, a study could investigate how the experience of HD might differ amongst residents of the United States of America, or Europe, or how the experience might be different for individuals who reside in rural areas of Canada. Each of these populations may be exposed to different services and have support communities that are different from those described within this thesis.

**Conclusion**

The majority of social science research on HD has focused on predictive genetic testing (e.g., Bombard et al., 2007; 2008; Cox, 1999; 2002; 2003; Cox & McKellin, 1999). This thesis has explored several of the salient issues outside of this test, specifically in relation to attitudes towards death and suicide, experiences with psychiatry, interactions with health services and bureaucracies, and the needs and experiences of caregivers. This work has also sought to respond to Wexler's (2008) critique that the documented experience of HD is dominated by the accounts of doctors and clinical researchers, by documenting these experiences from the perspective of affected individuals and their informal caregivers. It is my hope that this thesis will be just one of many forays into these aspects of HD and that the experiences of individuals after genetic testing will be as competently described as they have been vis-à-vis testing.


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Appendix 1: Methodology

A Note on the Original Aims of this Thesis

Before I describe the relevant methodological issues of this study, it is important to first touch upon its original aims. Initially, this study focused on two diseases, HD and Amyotrophic Lateral Sclerosis (ALS), also known as Lou Gehrig’s Disease. The thesis was to be based around questions of both time and embodiment. For instance, I was originally hoping to contrast how individuals with HD and ALS conceptualized dying, as the former is terminal in approximately 15 years, while the latter is terminal in two. Specifically, I was interested in the narratives that both cohorts would have about their diagnosis. This narrative analysis was intended to be presented alongside the thematic, grounded theory analysis.

I was also particularly interested in how these two cohorts would conceptualize “mind” and “body”. As has been discussed already, HD affects individuals comprehensively. However, ALS degenerates the body while leaving the mind relatively intact. Although the Cartesian division between mind and body is often portrayed as passé within sociological literature, I was interested in how participants would orientate to it. I was also interested what their accounts might say about Merleau-Ponty (1962) and Frank’s (1991; 1995) discussions of the importance of embodied experience.

Unfortunately, these questions remained unanswered. Getting access to the HD community (described below) was, as far as research goes, exceptionally easy. In contrast, my experience with the ALS community was rather nightmarish. I spent five months in the laborious process of obtaining ethics at a local hospital in an attempt to recruit. Despite my best efforts, I was unable to obtain a single participant. Given time constraints, I made the decision to switch
my thesis from a comparative study, to HD-centric. This decision had clear ramifications for analysis, as I was unable to address several of my original research questions.

It would be unfair for me to offer an explanation of the differences in accessing these two populations. Indeed, if I had taken different steps, such as interviewing ALS healthcare providers, I may have had a better outcome. However, it is still important to describe the original aims of this project and the difficulties that were encountered that necessitated the project’s alteration. Having described this transition from comparative analysis to an HD-centric investigation, I will now focus exclusively on the methodological issues relevant to my study of HD.

**Getting Access and Recruitment**

After gaining approval from the UBC Behavioral Research Ethics Board (see Appendix 6), my advisory committee and the Department of Sociology, recruitment of potential respondents began. First, permission was requested from the HDS to place flyers (see Appendix 5) at events scheduled for their members and to disseminate information about the study on their mailing list. Second, my personal contacts with the British Columbia Centre for Disease Control were accessed to contact both possible participants and potential gatekeepers. Third, the UBC’s HDRC was contacted for permission to place flyers within their facility on the UBC campus. Lastly, participants’ personal networks were used for recruitment (snowball sampling), with participants being encouraged to forward study details to friends and associates who may also have interest in participation. All participants were also offered a $20.00 honorarium, both to acknowledge their contribution and to provide an incentive to participate.

Although all of these avenues were pursued, only the HDRC, and to a lesser extent snowball sampling, were fruitful. The HSC referred the researcher to the HDRC. My professional network also proved to be of little use, with exception of Dr. McKellin (a committee
member on this study), who similarly suggested that the author contact a social worker at the HDRC. Thus, these various strategies for recruitment all led to the same individual at the HDRC.

The HDRC is funded by the HSC and is physically beside the HDMC at UBC. For the past 20 years, the HDRC has been staffed by a social worker (hereafter referred to as Ms. Tolley), who is part of the group of professionals that people with HD visit as part of their appointment at UBC, whether for diagnostic purposes or for an evaluation of progression. In this capacity, Ms. Tolley works alongside geneticists, neurologists, genetic counselors, and a psychiatrist, and has a portfolio that contains more than 400 clients (S. Tolley, personal communication, June, 6th 2009). Given that HD only affects approximately 6000 Canadians and that the professionals at the medical centre are consultants, meaning that no individual with HD is obligated to visit them, this is quite a significant number. Additionally, until recently, Ms. Tolley organized a bi-weekly support group for individuals with HD. She therefore has access to hundreds of individuals with HD, as well as having established solid rapport with her client base.

After contacting Ms. Tolley about the study, I scheduled a meeting with her to discuss potential recruitment. Ms. Tolley was quite enthusiastic about the study and agreed to distribute recruitment flyers at her support group. In addition, she also contacted several individuals she believed would be interested in the study by phone. Accordingly, Ms. Tolley was an essential resource to this study, which may have proved unfeasible without her support. Ms. Tolley also placed me in contact with a social worker on Vancouver Island that aided in similar recruitment strategies.

Individuals over the age of 19 were recruited into the study as long as they met one of two criteria: 1) they were gene-positive, or had been diagnosed with HD or 2) they were the caregiver of an individual with HD. That said, Ms. Tolley and I communicated regularly to ensure that participants were drawn from a diverse set of circumstances and stages within the disease trajectory (see sample section below).
In addition, I also recruited individuals via snowball sampling. For instance, when an individual with HD was interviewed, they were asked if an applicable caregiver, friend, or family member would also be interested in an interview. However, snowball sampling was deliberately limited due to its tendency to produce individuals of similar characteristics (Weiss, 1994).

While these strategies were essential for accessing participants within a feasible timeframe, all of the study participants were associated, in one-way or another, with the HDRC. Individuals who did not access these services, or were not aware of them, did not have an opportunity to be included in the study. As a consequence, this study's cohort receives comparatively more support than individuals who would not have had an opportunity to participate. This feature of sampling may have coloured the responses in three ways. First, the positive evaluations of HD-specific services communicated by respondents may not be representative of the larger HD community. Second, as these individuals are not receiving HD services, they may be experiencing greater burden. Third, their evaluations of health services may be more negative, as their avoidance of these services may have been caused by a particularly disturbing encounter with a health professional.

The optional use of the HDRC (as, once again, it is consultation service only) is also important in that participation in the study was not connected to any mandatory healthcare structures or distribution of healthcare benefits, thus potentially allaying participants' feelings of obligation to participate. However, as many participants were contacted by individuals involved in their care, additionally steps were taken to reduce feelings of obligation. Accordingly, I regularly reminded participants that I was not affiliated with the HDRC, HSC or HDMC and that they were free to speak about those services anonymously. Indeed, based on comments from Ms. Tolley and study participants, it appeared that if there was a motivating factor for some participants it was the offer of the $20.00 honorarium, as they are commonly asked to participate in research but rarely offered compensation. Respondents were also given the contact
information for my thesis supervisor and UBC Office of Research Services, should they have any concerns about their participation or the questions they were asked.

Data Collection

Qualitative in-depth, semi-structured interviews were the primary source of data collection for this project (Morse & Field, 1995). After being contacted, individuals were asked to schedule an interview at a time and place of their choosing. Four individuals lived outside a feasible area of travel and were interviewed over the telephone. Three of these individuals resided in remote areas of Vancouver Island and the other resided in the Interior of British Columbia. Importantly, several researchers suggest that telephone interviews yield similar accounts to face-to-face interviews (e.g., Midanik, Hines, Greenfield & Rogers, 1999; Sturges & Hanrahan, 2004). The remaining 26 interviews were conducted in person. Twenty-five of these interviews took place within the individual’s personal residence. The lone exception took place at a diner in Vancouver, due to the participant’s lack of privacy within her residence.

All interviews were conducted in complete privacy. However, multiple interviews took place near other people. For instance, in addition to the interview at the diner, one participant had numerous housemates (who did not know of her illness) pass through the interview area during the interview. In these instances I would change the topic to a pre-established subject (e.g., tourist areas in Vancouver) as to not disclose any sensitive information. That said, the amount of time during an interview where other individuals were within earshot was extremely minimal.

Interviews began with an explanation of the study, an invitation to ask questions and a recitation of the study procedures and ethical protocols. At this time, I also explained that my digital recording devices would be used to record the entire interview. One individual did state at this juncture that he wished for the digital recorder to be stopped during a particular topic, although he gave me permission to paraphrase and take notes on what was discussed during this
After the interview-interviewee contract was established, I secured written consent of the participant (see Appendix 3). Next, I delivered the $20.00 honorarium, which was offered to every study participant. Those individuals who were interviewed over the telephone both signed the consent form and were paid the honorarium in advance of the scheduled interview. All interviews were recorded using two Olympus DS-2 digital recording devices (one of which was a backup), with telephone interviews being facilitated by an additional piece of recording hardware.

Interviews ranged in duration from a half hour to two and a half hours. However, the vast majority of interviews lasted between one and two hours. Interviews were guided by an interview schedule (see Appendix 4) that emphasized descriptive questions that focused on the “hows” and the “whats” of the individual’s illness experience (Charmaz & Olesen, 1997; Emerson et al., 1995; Morse & Field, 1995). The project was based around four major areas of interest, or research questions. First, what are the experiences evoked by HD, particularly in lieu of the most pronounced physiological affects? Second, given the long period between diagnosis and palliation, how do individuals (re)-orient their lives? Third, what transitions occur for individuals who are the caregivers to those with such a disease? Lastly, given that lack of research that engages individuals with HD, what are their perspectives on current medical services? That said, as this was largely an inductive and exploratory work, the interviews were quite malleable, and whilst each area of interest was addressed, the interviews were more directed by the participant’s responses than the pre-established guide. Indeed, attention to participant directed issues produced the data forming the basis of the second chapter, as questions around psychiatry were not contained in the interview guide.

With just one exception, both members of a dyad were interviewed in the same day, one after the other, with the participants deciding on the sequence. While I preferred to conduct only one interview per day, this deviation was conducted so as to limit the time-burden placed upon
the dyad. Indeed, I often interviewed the caregiver before they proceeded to spend the duration of their partner’s interview making dinner or performing other household duties.

Upon completion of the interview, participants were again provided the opportunity to ask me any questions or talk about other relevant issues. At this juncture, almost all of the caregivers (N = 9) asked me why I was studying HD. Interestingly, only two individuals with HD asked a similar question. Upon answering any questions, I asked participants if they would be interested in a lay summary of the study’s findings, which all 30 participants requested.

After departing the site of the interview, I wrote detailed fieldnotes, which included descriptions of the interview location, relevant participant behaviour, basic descriptions of the participants themselves and researcher-participant interactions (Emerson et al., 1995). These fieldnotes served to add nuance to the project and provided another source of data for the subsequent analysis. In particular, the researcher was sensitive to the so-called “doorknob effect” whereby individuals state important information once the recorders are off and the researcher is on the way out the door (Warren et al., 2003).

All digital files were then downloaded onto a password-protected computer that was only accessible to myself. All consent forms were placed in envelopes and sealed in a locked filing cabinet. The demographic information was inputted into an Excel spreadsheet that was matched to numerical participant identifier (e.g., HD.1 or HD.2). Information such as residence, e-mail or telephone number was not included within this file.

The digital audio files, which were also coded alphanumerically, were then transcribed, checked for accuracy and had all identifying information removed or altered. Before data analysis, each participant was assigned a pseudonym. Originally, pseudonyms were created from the participants’ actual initials, so that “Michael Halpin” would become, for example, “Mark Harrison”. However, given the size of HD community, as well as the universal participant interest in the study findings, random names were chosen for each participant using an online
name generator (http://www.kleimo.com/random/name.cfm) that I then matched to the participant’s numerical identifier. As such, no document contains both the real name of participants and their pseudonym. The renamed interview files and fieldnotes were then uploaded into the Nvivo™ 7 software for qualitative data management in anticipation of data analysis.

Sample

Individuals Living with HD

Twenty of the study’s 30 participants were individuals living with HD. Three of these individuals were “gene-positive”. That is, although they had received predictive genetic testing that confirmed that they would develop HD within the course of their lifespan, they had not been diagnosed with symptom onset. Two of these three participants were also sisters, and are the only related members of the HD cohort.

Two of the participants were receiving some form of managed care at the time of the interview. The first was a 75-year-old man who spent the majority of his time residing in a care home in suburban Vancouver. This participant’s partner moved him into a care home once she was no longer able to care for him on her own, although he regularly visited her at their former residence. The second individual was a 24-year-old man who lived in a private care facility, also in a suburb of Vancouver. This individual had spent a year in psychiatric care after his diagnosis of HD before being transitioned into his current residence. Both of this man’s parents were absent in his life and his grandmother provided the financial support for his care. In contrast to the large care facility of the first participant, this individual resided in a house owned by his professional caregiver and housed two other individuals with chronic illnesses.

The rest of the 15 participants occupied a place on the HD trajectory between these two groups. All had received a neurological diagnosis confirming symptom onset; however, they all
still resided at their personal residence and either received support from an informal caregiver (n=9) or were managing their own care (n=7). As has been noted previously, HD has three major areas of effect (psychiatric, cognitive and motor) and individual’s experience can vary substantially. Accordingly, there was great diversity between individuals in regards to the symptoms that they reported experiencing. For instance, one 76-year-old male participant had no experience with the psychiatric or motor aspects of HD. However, his cognitive abilities, particularly his ability to multi-task, organize and remember had been affected by the illness. In contrast, a 36-year-old female participant experienced symptoms in all three main areas. Thus it is important to note that the age of the participants or their distance from diagnosis does not correlate with their symptoms. Although, generally speaking, the best gauge for symptom progression and severity is the individual’s CAG count, current research suggests that the CAG count is only an accurate measure when the number is particularly low (i.e., 37-42) or particularly high (i.e., 50+) (Benjamin et al., 1994; Rosenblatt, 1999). The majority of individuals with HD occupy the space between these limits and, as such, it is difficult to determine how they may experience the disease over time. Perhaps representative of the ambiguity around this number was the fact that the vast majority of participants had no knowledge of their CAG count.8

Regionally, the participants were drawn from various areas of British Columbia, although all participants resided in the southern portion of the province. In total, 13 individuals were from the Greater Vancouver Regional District, two individuals were from the outlying areas of Vancouver, four individuals were from Vancouver Island, and one individual was from the Interior of British Columbia. A major limiting factor on participants’ locations was the desire to have the majority of interviews conducted in-person. Financial and logistical constraints resulted

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8 Interestingly, this lack of interest in the CAG count is vastly different from my previous experience with individual’s engagement with medical numbers. For instance, the prostate specific antigen score was a ubiquitous topic in prostate cancer support groups.
in the majority of participants being recruited near the two major urban areas in British Columbia, Vancouver and Victoria.

As suggested above, the ages of the participants varied quite considerably, from 23 to 83 (M = 54). The mean age of 54 is quite reflective of the average age of individuals experiencing HD (Paulsen, 2004). However, there were two individuals, one aged 23 and the other 24, who are of exceptionally young age for HD. As has been mentioned previously, there is another category of HD called JHD, which affects individuals in their teens. Despite ongoing speculation about the 24-year-old individual at the HDMC, as a result of his sudden symptom onset of after a drug overdose, neither of these individuals definitively meet the criteria for JHD, and, as such, age wise, they are outliers in comparison to a typical HD cohort.

In terms of the gender, 12 of the individuals were male and eight were female. While there is a popular misconception, repeated by several of this study’s participants, that males have an increased propensity to develop the disease, HD affects both sexes equally. However, the Cytosine-Adenine-Guanine (CAG) counts do become more “unstable” when inherited from the father’s side; that is, the number of repeats can grow dramatically in the next generation (Goldberg, et al., 1993). Therefore, a son or daughter inheriting the condition from their father can experience a dramatic increase in their CAG count compared to their father’s, and may even experience symptoms before they present in their father (Rosenblatt, et al. 1999), which may have implications for the disease’s progression and severity. In contrast, when inherited from the mother, there is typically no significant variation in the CAG count. This gender difference is still being investigated. However, as the disease itself still affects men and women equally, an attempt was made to recruit similar numbers of male and female respondents.

Distance from diagnosis ranged from 16 to less than one year (M= 6). Once again, time since diagnosis did not appear to relate to experience of symptoms. For instance, the 24-year-old residing in a care home had only been diagnosed in 2006, whilst a woman diagnosed in 1997 was
still living on her own. Indeed, as can be noted in the diagnosis date range, one participant (who is still quite healthy) had been diagnosed 16 years ago, which is a year greater than the high end of the average life expectancy range post-diagnosis.

Only four of the participants were still employed. Furthermore, two of these individuals were the sisters who had not received a diagnosis of HD. The individuals who had received a diagnosis of HD were experiencing difficulties and apprehension at their jobs. For instance, one 23-year-old woman was routinely mocked by her co-workers for her inability to complete simple tasks. While the individuals who had ceased working did collect health and employment benefits, many of them experienced similar occurrences towards the end of their careers, with many being encouraged to take earlier retirement or being terminated. When working, the participants were employed in diverse settings, such as carpentry, truck driving, banking, shipping and dentistry. The incomes of these professions vary considerably; however, only three participants reported imminent financial stress. This situation may change, however, as the severity of the disease advances; it is important to note that the two individuals in managed care had an abundance of financial resources at their disposal.

Ethnically, the participants were quite homogenous. The entire HD cohort ethnically identified as being of either Canadian or Western European (e.g., English, German, Dutch) origin, with the exception of one participant who identified as Jewish. Importantly, this is not reflective of the epidemiology of HD, as all ethnic groups, with exception of individuals originating from Finland and Japan, experience the same approximate rates of the disease (Harper, 1991). However, this is reflective of the clientele of the HDRC, which only has two families (one Asian and one African) that do not identify as North American or Northern European (C. Power, personal communication, June 7th 2009).

In terms of marital status, 12 participants were either married or common-law, two were divorced, two were single, and four were widows. While obviously not a representative sample,
this contrasts research on martial status and chronic illness that suggests that women with chronic illness are divorced at higher rates then men with similar illnesses (Charmaz, 1993). The male divorcée did not disclose the nature of his divorce, and the female divorcée within the cohort was divorced after her husband began to suspect that she had a serious illness. In contrast, both one male participant and one female participant began dating, and then subsequently married their respective partners, well after they had been diagnosed with HD, a fact that was disclosed before their romantic relationships began. Of the 12 individuals in dyads, seven had partners that also participated in this study, which will be elaborated on in the subsequent section.

Caregivers

Ten of the study’s 30 participants were informal caregivers. The majority of these individuals were spouses (n=9) and one individual was a grandmother. However, many of these individuals were dealing with HD in numerous family relations. For instance, one participant was caring for her husband, son and granddaughter. As mentioned above, seven of the 10 participants were spouses of individuals in the HD cohort. In two of the three cases where the caregiver’s partner did not wish to be interviewed it was due to the severity of their symptoms, while, in the third case, it was due to the individual not believing he had HD. An eleventh individual, a professional care provider, was also interviewed. Although a professional, this individual was described as being more of a friend to the individual he was caring for. However, his experiences and perspective were substantially dissimilar to the other caregivers. For instance, while the majority of caregivers worried about the economic toll caused by HD, this individual’s income was derived from HD care. He was also connected to the person that he was caring for, but they did not have an emotional bond comparable to other participants. Subsequently, he was not dealing with the same feelings of loss, grief and isolation as the other caregivers. While this individuals
experience is still very important, it would only have been appropriate to utilize if the study sampled more professionals.

The caregivers' ages were more homogenous than those of the HD cohort, ranging from 37 to 65 (M= 54). This is perhaps unsurprising, as the two youngest individuals with HD were single and reported difficulties in negotiating romantic relationships. The caregiver cohort also included two newlyweds, both of whom knew of their partner's HD before marriage; one of these newlyweds discussed how the decision to maintain the relationship was quite difficult. For the rest of the caregivers, HD appeared well into their relationships.

With regards to gender division, nine of the 10 caregivers were female. Male partners did not offer explanations for their lack of interest in the study and were difficult to get a hold of to discuss potential interviews. Also, only four (of eight) women within the sample had partners at the time of our interview. The lone male caregiver to be interviewed noted that he did not often see other men at events, gatherings or support groups. When the author attended HD events in an informal capacity, individuals often talked about this caregiver in terms of their concern that he would leave his wife or how "good" he was for staying with her. These topics were not discussed vis-à-vis women who stayed with their affected husbands.

Six of the 10 caregivers were currently employed, with types of employment including realty, sales, printing, and business ownership. Many of these individuals worked full-time positions and then transitioned into a caregiving role once they returned home. Several individuals in the study were recently retired and suggested these dual-occupations were part of their motivation for retirement.

Like the HD cohort, the majority of caregivers ethnically identified as North American or Northern European (n=9), with one individual identifying as Punjabi. For the spouses, this similarity to their partners is reflective of social research on marriage trends, which suggest that people tend to marry those of similar backgrounds (Kalbach, 1991). The Punjabi participant
knew of her partner’s HD before they began dating and stated that her culture and her role within her family kept his illness from being perceived as a reason to not pursue a relationship.

Data Analysis

Analysis and the initial research questions, were framed by a number of works within the fields of philosophy, medical sociology and medical anthropology. A number of philosophical works on death and meaning, such Nietzsche (1977; 2000), Kierkegaard (1989) and Frankle (1984), were particularly influential on the overall focus of this thesis and, while not explicitly cited, also framed the first chapter. Conceptualizations of the body and illness, specifically Frank (1991; 1995) and Merleau-Ponty (1962), were also critical in orientating this work to the experiences of the ill. Although their work takes place in different contexts, Kleinman (1978), Farmer (2001) and Briggs and Mantini-Briggs (2003) were similarly influential in the decision to ground analysis in the accounts of individuals experiencing illness.

Data analysis also drew on numerous aspects of grounded theory, with particular attention to its uses in previous health research (e.g., Bottorff et al., 2006; Glaser & Strauss, 1964; 1965; 1968; Johnson et al., 2008). Specifically, line-by-line, thematic and case comparison analyses were utilized. Interviews and fieldnotes were coded and analyzed, both to increase the depth of the analysis and to compare each theme across multiple data sources.

Line-by-line analysis was conducted with each individual transcript, with the author writing analytical memos and notes (Strauss & Corbin, 1998). The transcripts were then re-read, with the analytical notes being drawn into larger codes that were then used to more efficiently to organize the data (Strauss & Corbin, 1998). These broad codes formed the basis of the coding structure used within the NVivo™ software program. Coding runs were then generated from these broad codes and each was read multiple times to develop emergent and inductive themes, which form the basis for this study’s findings.
In addition to the line-by-line and thematic analyses, case comparison analysis was also used to provide an extra analytical perspective. This stage of analysis utilized the Microsoft Excel spreadsheet program to compare individual cases (e.g., an individual with HD or a caregiver) across the entire cohort sample (Miles & Huberman, 1994). This technique allowed the data to be responsive to the particulars of an individual case, while also presenting it in relation to aggregate trends, subsequently increasing the study's ability to describe larger patterns and themes (Miles & Huberman, 1994). This approach also enabled the analysis to be reflective and representative of individual participants, whilst simultaneously not obscuring the analysis of the entire data set and presenting the whole as configurations of parts (Ragin, 1987). Importantly, this strategy also inhibited the privileging of data drawn from the thematic analysis by anchoring participant statements within their individual context.

During both stages of analysis (line-by-line/thematic and case comparison), codes were not considered to be mutually exclusive. As such, items could be coded under multiple headings and appear in the coding runs that generated different themes. However, once a quote or description was used within one section of the project findings, it was not used as evidence within another.

While analysis was loosely guided by research questions that informed the interview guide (e.g., what are the experiences evoked by HD, particularly in lieu of the most pronounced physiological affects?), and orientated towards phenomenology and existentialism (see above), it was not informed by any particular theoretical perspective or paradigm. Rather, as is suggested by numerous researchers (e.g., Glaser & Strauss, 1967; Latour, 2005), theories were utilized when they were beneficial for the analysis at hand. As such, the study utilized the works of individuals like Bakhtin, Said and Foucault where useful, but no single theorist monopolized the analysis.
Finally, given the lack of literature on a number of aspects of HD experience, a lay summary of study findings will be made available to the participants. As mentioned previously, the entire study cohort, as well as several care professionals, have expressed interest in such a summary. Before any of the projects findings are considered for potential publication, reflections, opinions and responses from these individuals will be solicited, thus providing a form of member checking on the data (Creswell & Miller, 2000).

**Participant-Researcher Interactions**

Throughout the project, there were several significant participant-researcher interactions. While one of these issues, reactions to discussing death and dying, was anticipated, several others were not. These issues included my progressive involvement with the HD community and with specific individuals in that community.

It was anticipated that discussing death and dying might be difficult for the participants. As such, although my own experience, as well as qualitative literature (e.g., Frank, 1995; Rapport, 2007), suggests that such conversations are often beneficial for the participant, a great deal of planning and preparation went into how issues of dying were to be discussed. Although participants were interviewed in regards to their overall experiences with HD, they were also informed that one of the areas of investigation would be issues relating to death and dying. Participants were also reminded before the interview commenced that they could stop the interview at any time, as well as refuse to answer any specific questions. To minimize any distress the topics may cause, questions related to death were placed in the middle of the interview guide, such that these questions occurred after rapport had been established, but are were far enough from the conclusion of the interview to ensure a successful transition to more mundane topics. Furthermore, as numerous methodologists have suggested, the topic was initially raised by asking a general question about the participant’s previous experience with
palliative illness, which then bridged into more specific questions about HD (Cox, 2004; Lofland & Lofland, 1995; Weiss, 1994). In case any individuals expressed distress over this topic, I also carried the nursing hotline number, which provides anonymous telephone access to registered nurses trained in dealing with medical and psychological issues.

Despite these measures, discussions on death raised apprehension for both parties. For instance, I frequently made jokes or light-hearted comments after the discussion had concluded in an attempt to alleviate my anxiety and normalize the conversation. Several participants began to cry during this section of the interview; however, no one accepted a request for a break in the interview or a change in topic. Indeed, despite the emotionality of the conversation, participants were exceptionally forthcoming and blunt during these conversations. One participant went so far as to suggest that the use of the word “palliative” was avoiding the topic and he much preferred the term “death” or “dying”. Another participant, a caregiver, expressed relief that she was able to talk about death, as she was unable to find a health professional who would discuss end of life issues with her. While discussing death often changed the tone of the interview, it also seemed to improve the rapport, which was perhaps best exemplified by one caregiver who joked that the interview felt like a “support group”. It should be noted, however, that the discussions in no way resembled a therapeutic interaction (see Weiss, 1994, for a discussion of the differences). Discussions about death and dying therefore provided somewhat of a paradoxical interaction. Both parties, experienced anxiety during this stage of the interview, but there was also a great amount of openness and relief.

I did experience one moment of acute anxiety following an interview were a participant (the project’s third recruit) discussed suicide. As alluded to previously, this individual gave me permission to report what he had said, but asked that the digital recorder be turned off during the period that suicide was discussed. Additionally, before the participant stated what he did not want recorded, he went over the terms of the consent form once more, to ensure that what he
stated would not be disclosed. After the interview, I experienced a great deal of distress and was questioning issues around breaching confidentiality. I first consulted the several applicable ethical policies (i.e., Tri-Council policy statement, CSA and ASA), but could only find a few vague statements that were applicable to situations involving potential self-harm. I then conferred with a mental health professional about issues relating to reporting of suicidal ideation and was informed that this case did not meet any of the three necessary criteria (immanent, clear and serious) for such an action (name withheld, personal communication, November 19th, 2008). The matter was further resolved during a follow-up phone call with the participant in question. Although the situation had a positive resolution, it still produced a great deal of stress and anxiety.

For myself, the most significant participant-researcher interaction was subconscious. Throughout the project, I experienced numerous dreams where I was an individual with HD. As the research progressed, I began to have dreams of being specific participants and displaying their particular symptoms. These dreams commonly occurred after I had spent the day analyzing transcripts, especially when the transcripts contained information that I already found particularly deflating. One transcript in particular, which recounted the experiences of a young woman, negatively affected me and was frequently set aside until it was the only transcript left to be coded or analyzed. During an informal conversation with a HDS volunteer, I asked how she was able to emotionally deal with the work that she did, in particular, how she was able to remain positive whilst seeing the people she knew deteriorate over the years? The individual responded that she received therapeutic counseling to deal with such issues. I was not engaged at a level comparable to this individual, but had certainly originally overlooked the personal psychological ramifications of the project.

In contrast to these experiences, there were also numerous occasions where I found myself being drawn into the HD community. All of these situations challenged my preference for
a clear differentiation between being a researcher and being a community member, which is a common anxiety for qualitative researchers (e.g., Duneier, 1999; Waters, 1999). Predictably, the first few instances were quite awkward. On the first occasion a participant invited me to his regular pub night, the second involved a younger participant giving me his Facebook address, which the participant also used as his business website. Neither of these invitations felt appropriate to accept and the process of declining them was very uncomfortable.

However, I eventually accepted invitations that blurred the line between member and researcher. Most notably, I volunteered at an annual HD camp for individuals with mild and moderate symptoms. Camp activities included physical activities—climbing wall, pool, and basketball—art and psychosocial therapeutic activities, such as guided imagery and group. In many ways, the camp was a source of discomfort. First, I debated requesting ethics permission to conduct ethnographic observations and eventually decided to be present only in the capacity of a volunteer. Second, I was assigned several individuals ("buddies") to supervise, with the corresponding duties often falling outside of my familiarity, particularly when I needed to aid one individual who had a seizure while in the swimming pool. That said, the camp, whether a source of formal observation or not, proved to be a pivotal learning experience and allowed me to interact with many of my former informants outside of the dynamics of a formal interview.

The camp also permitted me to hear to opinions of the support staff, which were dramatically different from those of the interviewees. Several members of the support staff had severe concerns about the resources and services that they were providing individuals within the HD community. In particular, I was told about a number of multiple distressing encounters between health professionals and individuals with HD. Indeed, I personally witnessed a particularly tense interaction over medication policies. These informal observations ultimately formed the basis for my earlier recommendations on areas of future research (see Chapter 5).
Since returning from the camp, I have received several e-mails from campers and have also attended a monthly lunch hosted by an individual with HD. Health professionals at the camp have also invited me on social outings, and have likewise suggested that I attend the bi-weekly HD support group, which I am considering after I finish my studies.

As a result of a previous research project that ended a friendship, I have recently leaned towards the perspective that participants and researchers should be separate entities. During this project, my attitude has been challenged in multiple ways, both by participants and other researchers. The researcher-participant relationship was particularly difficult during data analysis, as I worried about my abilities to present an “unbiased” picture, as well as feeling inauthentic when socializing with the people whom I was writing about.

As time has passed, I began to perceive my earlier preference for distance as slightly ridiculous. Foremost, I was already involved, committed and invested in the HD community. I frequently found myself feeling an increased sense of dedication after socializing with a former participant and the more that I got to know my participants, the more important it became to me to tell their stories. There certainly were difficult moments, but since data analysis has been completed, it feels much easier to engage with community members as a friend, rather than a sociology student.

In summary, I was well prepared for the possible discomfort that the project may cause for participants. However, I was considerably more naïve in regards my own sources of discomfort. This unease has been a beneficial learning experience and will also be an area that receives more thoughtful consideration in my future research projects. These issues were particularly salient during the process of writing. With writing completed, I found it much easier to engage the HD community and be myself.
Appendix 2: A Brief Note on the Canadian Context of Huntington Disease

Sir William Osler was the first Canadian to have a major influence on the understanding of HD. As mentioned above, Osler was both an enthusiastic fan of Huntington’s work and a highly regarded physician. Accordingly, he played a central role in the quick dissemination of Huntington’s results to the rest of the medical community (Harper, 2002; Wexler, 2008).

Aside from Osler, many of the HD developments within a Canadian context have taken place over the last four decades. In 1973, Ralph Walker, a teacher in Cambridge, Ontario began a small group for individuals affected by HD (Huntington Society of Canada, 2009). As this group grew, it eventually developed into the HSC. Starting as a small local group, the society expanded into a national association that connects individuals, supports research, distributes literature and raises awareness. One of the primary initiatives of the HSC was the creation of nine HDRCs. These sites provide resources and contact personnel for individuals with HD across Canada. Currently located within Kitchener, Ontario, the HSC continues to conduct similar work, despite a fairly limited staff and a restricted budget.

Most recently, the HDMC, located at the UBC Hospital, has been associated with several major HD research projects. First, their head physician, Dr. Michael Hayden, authored the most comprehensive history of HD (1981). Second, the HDMC team was narrowly beat out in the discovery of the HD gene that, as stated above, is integral in predictive testing for HD (S. Power, personal communication, June 2nd, 2009). Third, the team (in addition to numerous other UBC associates) authored a central policy paper on predictive genetic testing, which has been both used as a standard for performing HD predictive testing and as a template for similar policies regarding other diseases (Benjamin et al., 1994). Lastly, both the HDMC and the Vancouver HDRC have facilitated influential social science research on HD, particularly around issues of genetic testing.
Appendix 3: Consent Forms

THE UNIVERSITY OF BRITISH COLUMBIA

Consent Form
Study on Neurological Diseases

Principal Investigator: Michael Halpin, Masters Student in Sociology at University of British Columbia, Canada, (778) 229-6514, mahalpin@interchange.ubc.ca

Purpose: The purpose of this study is to understand the day-to-day interactions and activities of people living with neurological diseases, specifically Huntington’s Disease (HD) and Amyotrophic Lateral Sclerosis (ALS; also known as Lou Gehrig’s Disease). The study focuses on perspectives, experiences and quality of medical services. You are being asked to participate as an individual with a neurological disease. The results of the study will be analyzed and the results may be presented at conferences or be published in journals.

Study Procedures: The interview will take about 60-90 minutes. It will involve a conversation between you and the principal investigator (Michael Halpin) about your experiences. The interview will be recorded to capture the details accurately. The principal investigator will answer any questions you have about the procedures.

Confidentiality: Your identity will be kept strictly confidential. Recordings and documents will be identified only by code number and kept in a locked filing cabinet. Transcripts of the interviews, identified only with a code number, will be kept on the principal investigator’s computer and will be password-protected. Only the principal investigator will have access to the identities of study participants. You will not be identified by name in any reports of the completed study.

Risks/Benefits: The study will discuss your illness experience, which many individuals enjoy talking about. However, if you don’t feel comfortable answering any of the questions, you can simply tell the principal investigator and you don’t have to answer. Benefits of participating in this study include the opportunity to recount your experiences and giving feedback on healthcare services, which may be of use for individuals with similar illnesses. There are few studies on the experiences of individuals such as yourself, and your participation may help improve medical services.

Remuneration/Compensation: For participating in this study you will be offered an honorarium of $20.00 to thank you for your time.
Contact for information about the study: If you have any questions or would like further information about this study, you may contact the principal investigator Michael Halpin at (778) 229-6514 or mahalpin@interchange.ubc.ca. You may also contact the researcher’s supervisor, Dr. Zuberi: daniyal@interchange.ubc.ca.

Contact for concerns about the rights of research subjects: If you have any concerns or questions about your treatment or rights as a research subject, you may contact the Research Subject Information Line in the UBC Office of Research Services at (604) 822-8598.

Consent: Your participation in this study is entirely voluntary and you may refuse to participate or withdraw from the study at any time without penalty.

Your signature below indicates that you have received a copy of this consent form for your own records; and that you consent to participate in this study.

________________________________________
Subject Signature Date

________________________________________
Printed name of the subject signing above

________________________________________
Witness Signature Date
Consent Form

Study on Neurological Diseases

Principal Investigator: Michael Halpin, Masters Student in Sociology at University of British Columbia, Canada, (778) 229-6514, mhalpin@interchange.ubc.ca

Purpose: The purpose of this study is to understand the experiences of partners of individuals living with neurological diseases, specifically Huntington's Disease (HD) and Amyotrophic Lateral Sclerosis (ALS; also known as Lou Gehrig's Disease). The study focuses on caregiver experiences as well as quality of medical services. You are being asked to participate as the partner of an individual with a neurological disease. The results of the study will be analyzed and the results may be presented at conferences or be published in journals.

Study Procedures: The interview will take about 45-60 minutes. It will involve a conversation between you and the principal investigator (Michael Halpin) about your experiences. The interview will be recorded to capture the details accurately. The principal investigator will answer any questions you have about the procedures.

Confidentiality: Your identity will be kept strictly confidential. Recordings and documents will be identified only by code number and kept in a locked filing cabinet. Transcripts of the interviews, identified only with a code number, will be kept on the principal investigator's computer and will be password-protected. Only the principal investigator will have access to the identities of study participants. You will not be identified by name in any reports of the completed study.

Risks/Benefits: The study will discuss your experiences as a partner of an individual with a neurological illness. If you don’t feel comfortable answering any of the questions, you can simply tell the principal investigator and you don’t have to answer. Benefits of participating in this study include the opportunity to recount your experiences and giving feedback on healthcare services, which may be of use for individuals with experiences similar to your own. There are few studies on the experiences of individuals such as yourself, and your participation may help improve medical services.
**Remuneration/Compensation:** For participating in this study you will be offered an honorarium of $20.00 to thank you for your time.

**Contact for information about the study:** If you have any questions or would like further information about this study, you may contact the principal investigator Michael Halpin at (778) 229-6514 or mahalpin@interchange.ubc.ca. You may also contact the researcher’s supervisor, Dr. Zuberi: daniyal@interchange.ubc.ca.

**Contact for concerns about the rights of research subjects:** If you have any concerns or questions about your treatment or rights as a research subject, you may contact the Research Subject Information Line in the UBC Office of Research Services at (604) 822-8598.

**Consent:** Your participation in this study is entirely voluntary and you may refuse to participate or withdraw from the study at any time without penalty.

Your signature below indicates that you have received a copy of this consent form for your own records; and that you consent to participate in this study.

<table>
<thead>
<tr>
<th>Participant Signature</th>
<th>Date</th>
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Printed name of the subject signing above

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<tr>
<th>Witness Signature</th>
<th>Date</th>
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Appendix 4: Interview Guides

Interview Guide
Neurological Diseases Study
Michael Halpin
I. Demographic Information

We’ll begin with some background information about you.

1. How old are you?

2. How would you describe your ethnic background?

3. What is your marital status?

4. What is your occupation?

5. When were you diagnosed with your illness?

II.

6. In as much detail as possible, I’d like you to recount for me the day that you found out that you were ill.
   a. How was your illness confirmed?
   b. How long was the process of diagnosis?
   c. What symptoms were you experiencing at this time?

7. How did others, such as your family and friends, react to you afterwards?
   PROBE: What were some differences you noticed in your relations with them?

8. [If applicable] Without any major symptoms, what were your thoughts on being diagnosed with this disease?

III.

9. How has your diagnosis changed the way that you view your life?
   PROBE: Specifically, how have your plans for the future changed?

10. If you could tell the story of your life before your diagnosis, what would it be?
    PROBE: How has that story changed?

11. Overall, how do you think your diagnosis will impact your life?

IV.

12. [HD only] Some neurological diseases only affect cognition. How do you think those individuals experiences differ from your own?

13. [HD only] Other neurological diseases only affect movement. How do you think those individuals experiences differ from your own?
14. [HD only] How are genetic diseases different from other diseases? How does being diagnosed with a genetic disease change the way you think of your body?

15. [ALS only] Some neurological diseases also affect cognition. How do you think those individuals experiences differ from your own?

16. [ALS only] The cause of ALS is still being investigated, how does this affect the way you think about the disease?

17. Have you, or do you currently, have any other major health issues?
   a. If so, how are these similar to your neurological illness?
   b. How are they different?

18. What changes have you noticed in your body since diagnosis?
   a. How have these changes affected you?
   b. What future bodily changes do you expect?
   c. How have you prepared for these changes?

V.

19. What are the most important things for individuals who know nothing of your illness to learn?

20. How do you think your illness is depicted socially?
    PROBE: What does the average person expect your illness to be like?

21. Do you have previous experience with palliative illness (e.g., through the passing of a loved one)?
    a. How do individuals with you illness perceive dying differently from others?
    b. Have you made any preparations for the last stages of your illness?
    b. How has your diagnosis altered the way you think of death and dying?

22. How do individuals with your illness view life differently from others?

VI.

23. Could you tell me about your interactions with your primary health care professional?
    a. What other health professionals do you interact with during the course of your care and treatment?
       PROBE: How have these experiences been?

24. How effective are these services?
    a. How quickly are the services delivered?
       PROBE: Are there long waiting lines for the services you need?
    b. How would you rate the quality of these services?
       PROBE: How could these services be improved?
    c. How do the services you receive here compare to medical services you’ve received in other locations?
25. Could you tell me about the best experience that you’ve had with medical services? a. What was your worst experience?

26. What aspect of your illness receives the most support?

27. What aspect receives the least support?

28. What do medical professionals need to learn about your illness?

29. If you could recommend the creation of a new medical service for individuals with your illness, what would it be?

30. Are you a member of any support communities (e.g., support groups, online communities of societies)?
   a. [if yes] What has this experience been like for you? What are the positives and negatives?
   b. [if no] What are your reasons for not seeking out an organization like this?
   c. Outside of these formal groups, do you have contact with individuals who also have this illness? What are these interactions like?

VII. Wrap-up

We have almost completed the interview, I just have a few general questions for you.

31. Are there any important details or aspects of your experience that you think I’ve missed?

32. Are there any other issues that you would like to talk about?

33. Do you have any questions, comments or concerns about this interview?

34. Please feel free to contact me if you have any other questions or comments.
Partner Interview Guide
Neurological Diseases Study
Michael Halpin
I. Demographic Information

We'll begin with some background information about you.

1. How old are you?

2. How would you describe your ethnic background?

3. What is your marital status?

4. What is your occupation?

5. Around what date did you learn about your partner’s diagnosis?

II. Transition into Care-giving

6. In as much detail as possible, I would like you to recount for me the day that you found out that your partner was ill.

7. How did your partner’s diagnosis affect you personally?

8. When did you first notice symptoms of your partner’s neurological illness?
   a. How did you communicate about these symptoms with your partner?
   b. How did you communicate about these symptoms with others?

9. How has your partner’s diagnosis changed the way that you view your own life?

10. How are you currently involved in your partner’s care?
    a. What are the everyday caregiving activities you engage in?
    b. How did you transition into caregiving?
    c. How does your role as a caregiver compare to your previous roles in your relationship?

III. Differences between Diseases

11. [HD only] Some neurological diseases only affect cognition. How would being a partner in this context be different from your own experience?

12. [HD only] Other neurological diseases only affect movement. How would being a partner in this context be different from your own experience?

13. [HD only] How are genetic diseases different from other diseases?

14. [ALS only] Some neurological diseases also affect cognition. How would being a partner in this context be different from your own experience?

15. Have you previously been in a caregiver role with your partner?
    a. If so, how was this similar to your current situation?
    b. How was it different?
16. Has your partner previously been in the caregiver role while you experienced an illness?  
   a. If so, how was this similar to your current situation?  
   b. How was it different?  

IV. Perceptions of Medical Services  

17. Could you tell me about your interactions with your primary health care professional?  
   a. What other health professionals do you interact with during the course of your partner’s care and treatment?  
      PROBE: How have these experiences been?  

18. How effective are these services?  
   a. How quickly are the services delivered?  
      PROBE: Are there long waiting lines for the services you need?  
   b. How would you rate the quality of these services?  
      PROBE: How could these services be improved?  
   c. How do the services you receive here compare to medical services you’ve received in other locations?  

19. Could you tell me about the best experience that you’ve had with medical services?  
   a. What was your worst experience?  

20. What aspect of your partner’s illness receives the most support?  

21. What aspect receives the least support?  

22. What do medical professionals need to learn about your partner’s illness?  

23. If you could recommend the creation of a new medical service for individuals with your partner’s illness, what would it be?  
   a. If you could recommend the creation of a new medical service for partners of individuals with neurological illnesses, what would it be?  

24. Are you a member of any support communities (e.g., support groups, online communities of societies)?  
   a. [if yes] What has this experience been like for you? What are the positives and negatives?  
      b. [if no] What are your reasons for not seeking out an organization like this?  
   c. Outside of these formal groups, do you have contact with individuals who also have this illness? What are these interactions like?  
   d. Do you have any contact with other partners of individuals with neurological illnesses? What are these interactions like?  

VII. Wrap-up  

We have almost completed the interview, I just have a few general questions for you.
25. Are there any important details or aspects of your experience that you think I’ve missed?

26. Are there any other issues that you would like to talk about?

27. Do you have any questions, comments or concerns about this interview?

28. Please feel free to contact me if you have any other questions or comments.
Appendix 5: Recruitment Flyer

University of British Columbia
Department of Sociology
Neurological Illnesses Study

Participants are currently being recruited for a study on experiences with Neurological Diseases

Who can participate?
- Anyone with Huntington Disease who is over 19 years of age.

What is required?
- One 60-90 minute in-person interview
- The interview will cover issues of:
  - Diagnosis
  - Experiences with the effects of Huntington Disease
  - Talking to family and loved ones about Huntington Disease
  - Medical Services

Why should you participate?
- The opportunity to give feedback on the medical services that affect you
- All participants will receive copies of the study findings
- All participants will receive a $20 honorarium

For more details contact Michael Halpin
Phone: 778-229-6514
E-mail: mahalpin@interchange.ubc.ca
Appendix 6: University Ethics Certificate
CERTIFICATE OF APPROVAL - MINIMAL RISK

<table>
<thead>
<tr>
<th>PRINCIPAL INVESTIGATOR:</th>
<th>INSTITUTION / DEPARTMENT:</th>
<th>UBC BREB NUMBER:</th>
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<tr>
<td>Danial M Zuberi</td>
<td>UBC/Arts/Sociology</td>
<td>H08-02194</td>
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INSTITUTION(S) WHERE RESEARCH WILL BE CARRIED OUT:

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<tr>
<th>Institution</th>
<th>Site</th>
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<tr>
<td>UBC</td>
<td>Vancouver (excludes UBC Hospital)</td>
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Other locations where the research will be conducted:

In-depth interview will take place in a private location of the participants choosing, such as their personal residence.

CO-INVESTIGATOR(S):

Michael A. Halpin

SPONSORING AGENCIES:

Alberta Ministry of Advanced Education and Technology - "Neurological diseases and the construction of meaning"
Michael Smith Foundation for Health Research - "Neurological diseases and the construction of meaning"
Social Sciences and Humanities Research Council of Canada (SSHRC) - "Meaning in neurological diseases"

PROJECT TITLE:

Neurological Diseases: Time, Meaning and Narratives

CERTIFICATE EXPIRY DATE: October 1, 2009

DOCUMENTS INCLUDED IN THIS APPROVAL:

<table>
<thead>
<tr>
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<tbody>
<tr>
<td>Protocol</td>
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<tr>
<td>Proposal</td>
<td>October 1, 2008</td>
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<tr>
<td>Consent Forms:</td>
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<td>Consent Form</td>
<td>September 24, 2008</td>
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<td>Advertisements:</td>
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<td>ALS Flyer</td>
<td>September 24, 2008</td>
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<td>Huntington Flyer</td>
<td>September 24, 2008</td>
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<td>Questionnaire, Questionnaire Cover Letter, Tests:</td>
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<tr>
<td>Interview Guide</td>
<td>September 24, 2008</td>
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</table>

The application for ethical review and the document(s) listed above have been reviewed and the procedures were found to be acceptable on ethical grounds for research involving human subjects.

Approval is issued on behalf of the Behavioural Research Ethics Board and signed electronically by one of the following:

Dr. M. Judith Lynam, Chair
Dr. Ken Craig, Chair
Dr. Jim Rupert, Associate Chair
Dr. Laurie Ford, Associate Chair
Dr. Daniel Salhani, Associate Chair
Dr. Anita Ho, Associate Chair